mg., total protein, 29 mg. and chlorides 610 mg. per 100 cc. Results of Wassermann and gold curve tests for syphilis were negative.

The wound healed satisfactorily and the patient was discharged without further treatment.

When the patient was last observed, September 11, 1951, the symptoms were essentially the same as at the time of admittance three years before, and the meningocele did not appear to have increased in size.

DISCUSSION

The symptoms of anterior sacral meningocele are primarily owing to the mechanical effects of a pelvic tumor. Constipation and pain on defecation often are present. Serious obstetrical problems may arise because of partial occlusion of the birth canal. Occasionally minor nerve defects may be associated with the condition. Frequently symptoms are not pronounced and the tumor is found unexpectedly, as in the case presented here. Other congenital defects are often associated with this lesion.[3] Tumors of this kind occur more often in females than in males.

The diagnosis should be suspected when a tumor of the posterior pelvis is associated with a sacral defect. Sherman, Caylor and Long[5] observed that there were sacral defects in 23 of the 34 cases they reviewed. In many of the older reports, roentgen examination was not made and a sacral defect may have been present but overlooked. In the absence of a demonstrable sacral defect, the diagnosis must be confirmed by a myelogram or by aspiration of the tumor. It is important that aspiration or incision of the mass not be made through the rectum or vagina; meningitis developed in all of nine patients in the cases in which entrance was made by those routes, and seven of them died.[5] In the present case, unfamiliarity with the lesion was the reason for inability to make the proper diagnosis without surgical exploration and aspiration.

Surgical procedures performed in the diagnosis and treatment of these lesions have been accompanied by a high mortality rate. A course of "skillful neglect" has been advised, with the performance of cesarean section if sacral meningocele occurs as a complication of pregnancy.[5] Surgical ligation of the pedicle, which is often quite small, as it emerges through the sacral defect has been performed successfully several times and is the procedure of choice if it is decided that surgical removal is indicated.[1,2,4,6] The removal of the sac is optional since it does not secrete spinal fluid. The posterior approach to the lesion is attended with fewer hazards and complications. Since the patient in the present case had minimal symptoms which did not increase in three years, surgical excision was not carried out.

SUMMARY

A case of anterior sacral meningocele is presented.

The dangers associated with methods of approach that are not strictly aseptic, either for diagnosis or treatment of this rare lesion, are outlined.

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Scleromalacia Perforans

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DISEASES OF THE SCLERA are among the most serious diseases of the eye because of the complications which may occur.

Classification of the diseases of the sclera is made upon anatomic and clinical observations because knowledge of what causes them is limited. The case here reported was particularly hard to classify. It could not be placed in a textbook classification until several months after onset. Even then all the facts did not completely fit into place. The rarity of the various forms of scleritis makes it important that each case be thoroughly studied and reported, in the hope that sufficient information will be amassed in the literature to permit significant etiological conclusions.

Frequently observed is the disease entity nodular episcleritis,[1] and occasionally a form of superficial scleritis called by Fuchs episcleritis periodica fugax. The former is described as a chronic recurrent nodular inflammation of the episcleral tissue, usually bilateral. The latter and more regularly recurrent form is considerably more diffuse with edema and congestion of the tissues which is not present in the more benign cases. These superficial forms are not understood etiologically any better than the deep forms, but they rarely constitute a threat to the eye.

Deep scleritis has been divided clinically into anterior and posterior scleral inflammations. All cases of scleritis are chronic in character and in many one or more complications develop, such as sclerosing keratitis, and/or uveitis with sequelae such as anterior and posterior synechia, iris bombe, vitreous exudates, choroiditis, secondary glaucoma and panophthalmitis. Each complication may gravely threaten the sight and even the retention of the globe itself.

Posterior scleritis is usually associated with tenonitis and is most often secondary to retrobulbar infection extending from the sinuses. The chemosis and frequent concomitant occurrence of retrobulbar neuritis are so severe that not infrequently enucleation becomes necessary.

Anterior scleritis occurs as one of four separate clinical entities: Annular scleritis, scleroperikeratitis, brawny scleritis and scleromalacia perforans. Duke-Elder[1] described these diseases as separate and not necessarily related diseases. Indeed, each has definite characteristics not present in the others. These characteristics may be found, however, to be only the special reactivity of the area involved and probably have nothing whatever to do with etiologic factors. Proper reclassification of these diseases clinically must await further pathologic and physiological studies.

Annular scleritis is usually a diffuse induration of the circumference of the anterior segment of the sclera; that is, that portion of the globe in front of the equator. There are usually present pin-head sized, hard, white nodules in the sclera. Necrosis is unusual on a gross scale but it may be

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noted in microscopic examination of an enucleated eye. Scleritis of this type is frequently followed by or associated with deep infiltrations in the cornea, and when this occurs the disease is called scleroperikeratitis.

Brawny scleritis is a much more virulent disease clinically. The sclera, episclera and conjunctiva take on a brawny, gelatinous appearance. There is a true pitting edema of the globe. Never in a case of this type has degeneration been reported to have extended posteriorly beyond the equator. Histologically, degeneration is found in the sclera but nodules are not present and ulceration does not occur.

Scleromalacia perforans was first described as a specific disease by Van der Hoeve[9] in 1931, and in 1934[10] he reported four cases in which characteristic holes occurred in the sclera. These holes developed slowly. Occasionally a yellow nodule appeared which, upon puncture, exuded a yellow granular detritus. Three of the four cases occurred in association with polyarticular rheumatism.

Smoleroff[8] reported the presence of areas of focal necrosis of the sclera in three cases of rheumatoid arthritis.

Edstrom[2] reported a case in which flat grayish nodules occurred in the sclera. The nodules consisted of granulomatous tissue with necrotic connective tissue in the center. Results of culture and guinea pig inoculation of material from the nodules were negative for tuberculosis. The patient had severe arthritis, and the result of an agglutination test was positive for a hemolytic streptococcus.

Up to 1938 there had been 14 cases reported with several others of questionable classification. Verhoeff and King[11] added a case to this list and reviewed all the preceding cases from the available information. They formulated a composite clinical description as follows:

> The disease may occur in one or both eyes simultaneously or at intervals in patients in the 5th, 6th and 7th decades of life. Present usually are slightly elevated nodules which involve the sclera and overlying tissues. These nodules may occur in the sclera from the limbus to the equator. The congestion is most often only moderate and is limited to the nodules and their immediate vicinity. After several months the nodules regress, leaving shallow concavities. When the nodule is at the limbus it may perforate into the anterior chamber. Actual rupture of the globe is uncommon. Evidence of intraocular infection is slight unless purulent infection occurs. Posterior synechia or cataract may develop. In most cases rheumatoid arthritis exists at the onset of scleritis.

In most cases the disease is associated with rheumatoid arthritis, but not in all. Oast[7] reported a case which was identical clinically and pathologically with the case described by Verhoeff but in which there was no arthritis.

REPORT OF A CASE

A 59-year-old Mexican male whose right eye had become inflamed three weeks previously was first examined in June 1949. He was illiterate, lived out of town and returned irregularly to the outpatient department for treatment. Mild injection of the bulbar and palpebral conjunctiva of the right eye was noted. No discharge, follicles or papillae were observed and no preauricular adenopathy was present. There were a few pin-point areas of staining on the right cornea. The ocular media were all clear and both fundi were normal. The visual acuity was 20/70 in the right eye and 20/30 in the left. Each eye was correctible to 20/20 with concave lenses. No evidence of scleral disease was noted on gross or slit-lamp examination. No other significant abnormalities were observed in a complete physical examination.

The eye was treated with 30 per cent sulfacetamide solution locally for two weeks without improvement. After two weeks (five weeks after the right eye was affected) the left eye became involved. During the first week that both eyes were involved some episcleral injection developed in an area 6 to 8 mm. wide surrounding the whole circumference of the limbus in both eyes. The ocular media remained clear and the vision was not diminished. A tentative diagnosis of episcleritis was made and the patient was given salicylate therapy and local application of heat.

The patient returned to the clinic July 5 because of increased pain and tenderness in each eye. There was a 2 mm. by 4 mm. punched-out ulcer in the indurated conjunctival tissue at the 12 o'clock position in each eye. These ulcers had a yellow necrotic base and extended to the cornea. The patient stated that the ulcers had been preceded by yellow nodules. He was hospitalized for further study.

No new information regarding the eyes was elicited by detailed questioning. The patient had been in good general health until two years previously. During this two-year period he had received intermittent therapy by his family doctor for pain in the ankles, knees and shoulders. Salicylate treatment in large doses for arthritis had been given for two months prior to the onset of the eye disease, but had caused so much gastric upset that it had to be discontinued.

Except for the eyes, no abnormality was noted in physical examination. There was no swelling, tenderness or limitation of motion of the joints. Results of all laboratory tests at the time of admittance, including serologic tests for syphilis, were within normal limits. No evidence of disease was noted in an x-ray film of the chest. The uric acid content of the blood was 4.2 mg. per 100 cc. The reaction to a skin test with 0.01 mg. of old tuberculin was negative at the end of 72 hours, and to 0.1 mg. of the substance was 1 plus in 48 hours. The result of a Frei test was negative. Coagulase-negative staphylococci were noted in smears and cultures of material from the conjunctiva of each eye, but no abnormal cells were seen on smears. Normal conjunctiva covering a non-specific granulomatous process was observed in biopsic specimens taken from each ulcer.

X-ray films of the knees and ankles showed slight irregularity of both malleoli and some hypertrophic fringing, of long duration, of the margins of the articular surfaces of the tibial spines of both knees.

For two days after the patient was admitted to hospital the temperature was elevated one degree (Fahrenheit) at 4 p.m. but remained normal thereafter except when fever therapy was given.

The ulcers slowly became larger and deeper. The wounds made in excision of material for biopsy did not heal. The area of induration enlarged to 10 mm. back of the limbus in each eye. The patient was given 3.5 million units of penicillin, 30 gm. of streptomycin, and for four days was given therapeutic dosage of sulfadiazine in conjunction with local application of sulfathiazole and sulfacetamide, in succession or combination, without effect on the course of the disease.

When it became evident that the biopsy wounds were not healing, the ulcers were treated with a thermophore. Although this therapy stimulated some healing, it had to be discontinued because of a temporary rise in intraocular tension to 60 mm. of mercury (Schiotz).

After the patient had been under treatment for two months mild uveitis developed. It responded to foreign protein therapy with typhoid antigen, but the treatment had no effect on the scleritis. Operation to cover the ulcer in the right eye with a conjunctival flap was carried out. A specimen of diseased episcleral tissue was removed for guinea pig inoculation and the remaining episcleral tissue was cauterized with heat. A conjunctival flap was placed over

the entire ulceration and cornea of the right eye. Both eyes then began to improve. The patient was discharged from the hospital on November 1, 1949. No symptoms developed in the inoculated guinea pig, and when it was examined at the end of six weeks, no evidence of tuberculosis was noted.

A month after the patient was discharged, two firm, yellow scleral nodules which caused some discomfort appeared on the right eye, 10 mm. from the limbus. One was 3x6x1 mm. and the other 3x3x1 mm. The ulcer of the left eye was shallow and partially healed at the limbus. In January 1950, about six months after the onset of the condition, there was a recurrence of uveitis in both eyes. Reaction to a skin test at that time with 0.1 mg. of old tuberculin was again 1 plus in 48 hours. The uveitis improved and the patient was discharged after three weeks. In February the patient complained of pain in the chest and productive cough with some fever. In an x-ray film of the chest taken February 7 no evidence of active disease was noted. The patient was readmitted to the hospital in March, and an x-ray film then gave evidence of pneumonitis in the base of the right lung. The sedimentation rate was greatly accelerated. A culture was positive for Mycobacterium tuberculosis. The patient was transferred to a tuberculosis sanitarium in April. At that time both eyes were healing and the uveitis was inactive. The iris of each eye was bound down by many posterior synechiae. The scleral ulcer of the left eye which had not been covered by a flap was present but smaller. The scleral ulcer in the right eye had healed. Nodules were present in both eyes about 10 mm. off the limbus. In the right eye one nodule had healed to some extent, leaving the sclera thinned. The dark uvea could be seen through the yellow, semi-fluid center of the nodules. The patient died May 20, 1950, of pulmonary tuberculosis.

PATHOLOGY

In examination of specimens of tissue taken from the margin of an ulcer in this case of scleromalacia perforans it was noted that the conjunctival and episcleral tissue at the ulcer margin was in reality the edge of a nodule, the top of which had sloughed away.

The cellular debris in the ulcer margin consisted mainly of lymphocytes, polymorphonuclear leukocytes and plasma cells with nuclei in various stages of degeneration. The wall of the ulcer was composed of epithelioid cells infiltrated by many lymphocytes and plasma cells. Occasionally a plasma cell was seen to have undergone some colloid degeneration. In the wall of the ulcer were several giant cells having nuclear arrangement of Langerhans type. No histiocytes were observed in this section. There were fibroblasts surrounding and among the epithelioid cells but a notable lack of new vessels. The vessels near the margin of the ulcer were surrounded by small collections of lymphocytes and plasma cells. No actual thrombosis was noted in the section.

The features usually observed in examination of tissue in cases of scleromalacia perforans are (1) necrotic scleral nodules containing lymphocytes and degenerated collagen located in the anterior sclera; (2) epithelioid cells surrounding the necrotic center, often in radial arrangement; (3) giant cells of Langerhans type, but no macrophages.

DISCUSSION

Verhoeff made extensive histologic study of the lesion in the case reported by him. He described the lesion as a central mass of necrotic lymphocytes surrounded by five or six layers of radially arranged epithelioid cells. An occasional giant cell of Langerhans type was present, but there were no macrophages. Some eosinophils were present.

Prior to Verhoeff's exhaustive study, the only description of pathologic material was that it was either chronic inflammatory tissue or non-specific granulomatous tissue.

A case first reported by Kiehle in 1937[5] was described histologically in 1946.[6] The description closely paralleled that of Verhoeff, as did a case studied by Harbater in 1949.[4]

Eggers[3] reported similar microscopic observations in a case of what was termed *necroscleritis nodosa,* a name first advocated by Verhoeff[11] in 1938.

The case presented herein differed from the one first described by Van der Hoeve in that there was more diffuse episcleritis which spread downward to the sclera. In no previously reported case was there bilateral ulceration except where a nodule has been incised, and ulcers of that order usually remained indolent with little or no response to treatment. The sudden appearance of ulceration in the present case possibly represented obliterative periarteritis or endarteritis in the area involved, as that condition was frequently observed histopathologically, around the nodules.

The patient had hypertrophic arthritis but the absence of true rheumatoid arthritis in the case here reported is not distinctive. The occurrence of pulmonary tuberculosis, which was the ultimate cause of death, created a differential diagnostic problem. The eye could not be obtained for final complete pathological examination.

The similarity of the essential lesion of this disease to that of subcutaneous rheumatic nodule and to a tubercle must not be overlooked. The necrotic connective tissue and the cellular debris in the center surrounded by epithelioid cells is common to these three lesions.

The occurrence of ulcerative tuberculous scleritis or even a tuberculous process in the eye at all in the presence of active pulmonary tuberculosis is extremely uncommon. The facts that there was no response to streptomycin and that cultures and guinea pig inoculation were negative for tuberculosis were taken as presumptive evidence that these nodules and ulcerations were not primarily tuberculous. This was substantiated further by the biopsy of the ulcer margin, in which the conditions observed were not the pathologic changes associated with tuberculosis.

It is possible that the repeated use of fever therapy for the uveitis reactivated old quiescent pulmonary tuberculosis, but repeated tests and x-ray films gave no indication of pulmonary disease until the patient had been under treatment for eight months.

In several of the cases reported by other investigators cultures and animal inoculations were carried out, but in none was a tuberculous process demonstrated. Verhoeff stated that if the disease were of tuberculous origin, material from the lesion would be extremely likely to infect a guinea pig. He called attention to the fact that, although all the elements of a tubercle are present in many kinds of lesions, it is generally accepted that the cellular arrangement is the diagnostic feature. In none of the histologic material in any case was the typical eosinophilic center of epithelioid cells surrounded by lymphocytes of a typical tubercle observed.

There are more giant cells in scleromalacia perforans and not the characteristic palisading usually seen in a subcutaneous rheumatoid nodule. Possibly there is a slight alteration in the lesion because of some special reactivity of the scleral collagen. The proximity to the surface and consequent lower temperature may be of consequence. However, it is felt that the lesion is most probably a rheumatoid subcutaneous nodule in the eye.

In the tubercle, giant cells are produced in response to the presence of a fatty acid which is present in the organism. No source of this or any fatty acid is known in scleromalacia perforans at this time. Other chemical stimuli to

giant cell formation remain to be described. There may never be enough cases of scleromalacia perforans to permit as complete a study of the chemical factors as there has been in tuberculosis. Great strides have been taken in the past years in this field in other collagen disease, and inference from these studies may help answer questions about scleromalacia perforans. The response or lack of response to cortisone will certainly add to understanding of basic pathological chemical factors of the lesions. Cortisone was not available for the patient in the present case.

SUMMARY

A case is presented of scleral disease which clinically and pathologically resembled scleromalacia perforans, having scleral nodules progressing to indolent ulcers. The diagnosis was complicated by pulmonary tuberculosis. Local therapy appeared to be of value only in controlling secondary infection. Healing at the sites at which specimens for biopsy were excised from nodules was unsatisfactory; ulceration occurred and not until a conjunctival flap was placed over the wound did healing take place.

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Discussion by* A. R. IRVINE, JR., M.D., *Los Angeles

In recent years there has been considerable evidence that collagenous tissues often react violently to various antigens. This so-called hypersensitive state is particularly apparent in the scleral diseases described in the foregoing presentation. In addition to its high collagen content, the sclera is comparatively avascular. These two factors are primarily responsible for the clinical and pathological characteristics of scleral granulomatous disease. In this regard I have never been impressed with the distinguishing characteristics of the various types of clinical scleritis described by the author. It appears that the classification of keratitis as brawny, sclerosing, or as scleral malacia, is dependent upon a difference in degree and location rather than a difference in type of inflammation. Chronic granulomatous disease within the sclera produces multiple small foci of necrosis because of the avascular nature of the sclera, whereas a similar process in a tissue with a good blood supply might well develop to considerable size before necrosis occurs. The scleral pathologic changes encountered in the conditions discussed by the author depend upon the peculiar hyperreactive state and the histologic nature of the sclera, and have a wide variety of etiological agents.

Since the advent of cortisone, it has become extremely important to determine the specific cause in any given instance. Whereas many cases of so-called non-specific scleritis respond well to the use of cortisone, it is becoming evident that where bacteria are present the use of that drug is contraindicated. It appears clinically and experimentally that although cortisone suppresses the inflammatory response, it does not repress the growth of bacteria. Rabbits infected with tubercle bacilli will show apparent improvement and rapid regression of inflammation when treated with cortisone only to have massive necrotic lesions develop in which large numbers of tubercle bacilli are seen after a period of time in spite of continued cortisone therapy. The same is true of experimental syphilis. I have seen two instances of spontaneous perforation of disciform keratitis after prolonged treatment with cortisone drops. In another case a patient with chronic granulomatous scleratitis and uveitis became much worse and had perforation and atrophy of the involved eye after the use of cortisone topically and systemically. It becomes apparent, therefore, that it is not sufficient to classify scleral disease on the basis of its location and pathological anatomy, but that it is necessary to continue to search for the specific etiological agents.

Perineal Myoma

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TUMORS IN OR NEAR the prostate usually present no great diagnostic problem. Occasionally there may be clinical uncertainty as to whether a mass is primarily rectal or primarily prostatic. Most enlargements or masses in this area are owing to changes within the prostate—most frequently benign hyperplasia, malignant neoplasm, prostatic cyst, or prostatic abscess. Some masses deep in the perineum may arise as periurethral abscesses, others as perianal abscesses. Other lesions which may be felt by the examining finger as a mass in the region of the prostate are cysts of the seminal vesicle or of Cowper glands. Sarcoma may originate in or adjacent to the prostate and cause unusual rectal findings.

In the case here reported an unusual lesion raised a problem in the differential diagnosis of what might be called periprostatic tumors. In a careful survey of indexes of medical literature and various textbooks of urology and pathology no mention of a similar instance of perineal myoma was found.

CASE REPORT

A 41-year-old man was first observed May 20, 1949, because of complaints of urinary frequency, urethral irritation, feeling of resistance to emptying of the bowel or bladder and feeling of fullness in the perineum for the preceding month. These symptoms were first noted in association with non-specific urethritis.

Temperature was 99.2° F. Blood pressure was 230 mm. of mercury systolic and 140 mm. diastolic. The urethral meatus was hyperemic and contained a small drop of clear mucoid material. On rectal examination the prostate felt small and of normal consistency. Immediately above the prostate on the left side was a spheroid mass about an inch in diameter. It felt cystic.

Urine in both glasses of a two-glass test was cloudy and contained shreds of blood and mucus. The pH of the urine was 5.5, the specific gravity 1.018, and albumin content 3 plus. It contained no sugar. Upon microscopic examination