

they enlarge. They may be capillary or cavernous; both may bleed excessively and, as Watson & McCarthy (1940) put it, by the time the tissue is in the laboratory the fate of the patient is decided. Bleeding can occur following surgery or trauma or spontaneously if the tumour involves the gingivæ. Swelling is usually slow to develop and at first is bony hard. Pain is due to pressure on adjacent nerve elements (Stones 1962) and may be pulsating in nature. Radiology is an aid to diagnosis and Worth (1963) describes two principal radiographic appearances. In the first there is alteration in the structure of the bone, with coarse trabeculae radiating from the centre of the lesion like the spokes of a wheel, a well-defined border with normal bone and sometimes radiating spicules at the expanded periphery, which form the 'sunburst' appearance, described by Bucy & Capp (1930), resembling the sunray effect sometimes seen in sarcoma, except that the spicules tend to be shorter and the general bone density greater (Ingram 1964, personal communication). In the second and more common type there are cyst-like shadows which may be loculated or multiple giving a honeycomb or soap bubble pattern. Worth also mentions a diffuse type with poorly defined edges. In the present example there are features of both main types; coarse trabeculation but without the wheel-spoke arrangement, a buccal 'sunburst' and a cyst-like region in the ascending ramus. The partial loss of lamina dura and resorption of the teeth is stated by Smith (1959) to be characteristic; the neurovascular canal, not visible in this case, was reported to be enlarged in two previous cases (Battersby 1957, Dibble & Whelan 1962); carotid arteriography would clearly settle the diagnosis but is not without hazard (Cockett 1960) and should not be necessary.

Treatment of hæmangioma of the mandible can be divided into the emergency arrest of hæmorrhage when the lesion has been opened and the planned eradication of the non-bleeding lesion. In the former case Smith (1959) advocates concentration on local measures to obliterate the cavity with absorbable packing material under pressure rather than major vessel ligation which Broderick & Round (1933) and Brodsky (1934) reported not to affect the flow to any extent. For the other cases Smith states that radiation is the choice. Should surgery be undertaken, hypotensive anæsthesia may be of value in addition to other precautions to avert or arrest a hæmorrhagic crisis. No record was found in the literature of the use of sclerosing solutions in the treatment of mandibular hæmangioma.

Though it is uncommon in the jaws, hæmangioma should always be considered in the differential diagnosis of space-occupying lesions

of the mandible or maxilla; aspiration before biopsy or other surgery is advisable in doubtful cases. The fact that this case did not bleed excessively at biopsy was probably due to the fibrous stroma and low blood pressure in the spaces. Hæmangioma of the jaws may present varying clinical and radiological signs; therefore cases encountered in the future should continue to be reported to increase our knowledge of this lesion.

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The Syndrome of Jaw Cysts, Basal Cell Tumours and Skeletal Anomalies

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The syndrome of multiple cysts of the jaws, basal cell tumours of the skin and skeletal abnormalities is now well recognized; Gorlin *et al.* (1963) reviewed the main features and tried to assess the frequency with which the various abnormalities had been recorded.

As in all multi-lesional syndromes, mild or incomplete forms appear, so that it may be difficult to decide whether a case is the result of the syndrome or not and this may be important in order to assess the prognosis. Case 2 described here is mild but shows features which justify its inclusion. The main clinical features of the cases are as follows:

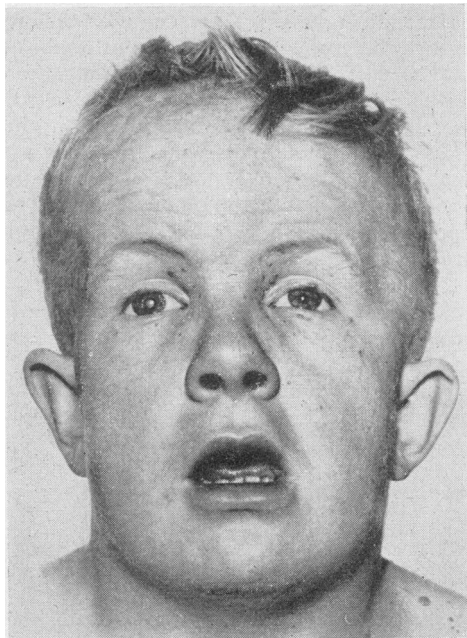


Fig 1 Case 1 This shows the parietal and temporal prominences, the broad nasal root, and basal cell naevi round the eyes

Case 1 Boy, now aged 14 (Figs 1, 2, 3)

This patient was found at the age of 1 to have severe hydrocephalus. A cerebellar astrocytoma was also found and removed. At the age of 2 'warts' were noticed on trunk and scalp. At 5 a cataract was seen in the right eye which is now blind, and five years later an opacity was noticed in the left eye. At 11, cysts apparently dentigerous were found in relation to all the second molars and to the upper canines. Two years later cysts had appeared in relation to the lower canines. In all, eight were removed. By the age of 13 the tumours on the back, now clearly basal cell carcinoma, up to 2 cm in diameter, demanded treatment. Wide excision and grafting were done.

This patient seems to have slight mental retardation and has a facies similar to those described by Gorlin *et al.* (1963). There is mild frontal bossing, a broad nasal root and well-separated eyes; the skull is large, with parietal bulging, calcification of the falx and an abnormally shaped sella turcica. The neck is very short and there is a bifid rib, with others broadened at the sternal end; there are abnormalities of the lumbar vertebrae and scoliosis. No other members of the family appear to be or to have been affected either by jaw cysts or multiple skin tumours.

Case 2 Girl, aged 11

This patient has dentigerous cysts in relation to all the second molars. She has a deformity of the

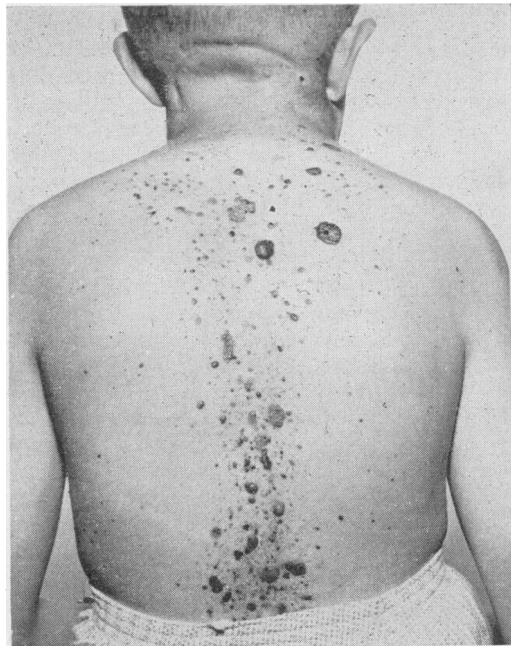


Fig 2 Case 1 The basal cell lesions on the back and shoulders are strikingly symmetrical in distribution

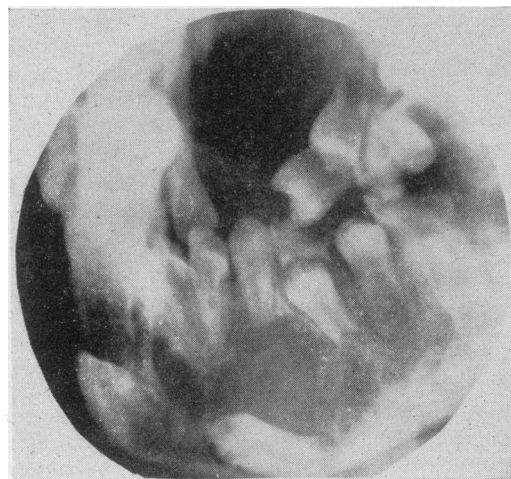


Fig 3 Case 1 This cyst, which extends across the midline to approximately the same distance on the opposite side, involves many teeth and is probably formed by fusion of several smaller cysts

chest producing an ill-defined prominence, about 10 cm in diameter, just to the left of the middle of the sternum and there are several bifid ribs. There are many skin lesions on the face especially round the eyes; these lesions are only a few millimetres in size and no biopsy has yet been carried out, though they appear to be enlarging slowly. The facies is rather similar to that described by

Gorlin *et al.*, having a broad nasal root, widely spaced eyes and mild mandibular prognathism. An epidermoid cyst was removed from the mid-line of the bridge of the nose at the age of 6. This patient appears to be of normal intelligence. The family history is uninformative.

Case 3 Male, aged 23

This patient had an epithelial lesion of the fifth finger, of unknown duration, removed at the age of 11; histologically it resembled a basal cell nœvus. At the age of 17 swelling of the jaws called attention to cysts and during the next two years five operations were performed to remove them. They were related to the upper and lower molars and to the lower incisors; at least two additional microcysts about 0.8 cm in diameter were also found at one of the operations.

The first basal cell carcinoma was removed from the eyelid at the age of 19 and he now has many of these tumours on the face, especially on and around the eyelids, up to about 0.7 cm in diameter. This patient has a facial appearance rather similar to Case 1 except that the tumours round the eyes are conspicuous.

There is calcification of the falx and an enlarged pituitary fossa; no other skeletal abnormalities were noted.

Histology

The cysts of the jaws from these patients show well-formed, stratified squamous epithelium of even thickness and with no inflammatory changes; the basal cell layer is well defined but there is no excessive proliferation of these cells. There is a suggestion of early keratinization in the main cysts while the microcysts from Case 3 are filled with keratin so that they resemble dermoid cysts.

The skin lesion in Cases 1 and 2 has the appearance of basal cell carcinoma; none resembles so-called epithelioma adenoides cysticum (tricho-epithelioma) as described by Binkley & Johnson (1951). They could therefore be regarded as basal cell nœvi (Lever 1961), i.e. basal cell lesions developing early in life but not showing deep invasion; active invasion may develop later, however, as Howell & Caro (1959) described and this is an additional reason for early excision.

Discussion

At the moment there is little knowledge of the nature of this disorder. Gorlin *et al.* (1963) consider there is evidence that the syndrome is transmitted as an autosomal dominant of poor penetrance or that there is partial trisomy of a small chromosomal segment. As to the pathology, there appears to be nothing characteristic about

the individual lesions in themselves. From the practical point of view it is important to recognize this syndrome; the patient is likely to have to face many operations for the cysts of the jaws, and more of these will probably appear later. In addition the skin tumours may be troublesome or disfiguring and require repeated excision; the eye lesions may have equally serious effects. The long-term prognosis is unknown but there seems to be a possibility, though not a strong one, that sarcomatous change may develop in the jaws.

It is interesting that 2 out of 3 patients presented here have an abnormality in size or form of the pituitary fossa; it is tempting to speculate whether or not this is also a characteristic feature of the syndrome.

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Hyperparathyroidism

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Miss C D, aged 60

Referred on 26.8.63 complaining of swelling in the anterior part of the mandible, which prevented her from having a lower denture.

The patient first noticed the swelling in April 1963, but did not seek treatment until July because she had severe pain in her back which was worse on climbing stairs and her dental practitioner's surgery was on the first floor. In July 1963 her remaining lower six anterior teeth were removed under local analgesia. The sockets healed well but the swelling continued to increase in size.

History: The pain in the back started in 1936, when she was 32, and had always been worst in the region of the right sacro-iliac joint. Radiographs of pelvis and lumbar spine in 1942 revealed nothing abnormal. The pain continued and in 1958 she retired from her employment as a school teacher on account of pain and discomfort in her lower back. She had noticed loss in height