

ment of port wine stains is becoming more effective with better graft techniques and the use of skin from donor sites with "blush" capabilities. 6) There is very little indication for the use of CO₂ snow, liquid nitrogen, or electrodesiccation. 7) Any form of irradiation is always contraindicated in the treatment of hemangiomas! 8) Lymphangiomas usually require staged surgical resection and reconstructions. They do not involute and portions of tissue containing the abnormal lymphatic network may be used in the reconstructions.

Comment

Hemangiomas have been defined and a classification has been proposed which combines the essential features of prognosis and optimum treatment methods. Treatment of each major variety of hemangioma has been outlined and special emphasis has been placed on the use of systemic steroid therapy as an invaluable aid in the management of the massive juvenile capillary-cavernous hemangiomas of infancy.

Some consideration has been given to the surgical treatment of port wine stains and other intradermal hemangiomas. Current approaches to the management of congenital arteriovenous fistulae are outlined.

Major complications of gamma and beta irradiation therapy are demonstrated to support the position that they are unwise and contraindicated methods of treatment for all forms of hemangiomas.

More attention should be paid to the psychic impacts of deformities resulting from hemangiomas. In that regard, plastic surgical techniques can be used more effectively and applied at younger ages than has been the custom in the past.

Several important distinctions have been made between lymphangiomas and hemangiomas in regard to prognosis and treatment.

References

1. Bennett, J. E. and Zook, E. G.: Treatment of Arteriovenous Fistulas in Cavernous Hemangiomas of Face by Muscle Embolization. *J. Plast. and Reconstruct. Surg.*, 50:84-87, 1972.
2. Brooks, B.: Discussion of Noland and Taylor. *Trans. South. Surg. Assoc.*, 43:176, 1931.
3. Brown, S. H., Neerhout, R. C. and Fonkalsrud, E. W.: Prednisone Therapy in the Management of Large Hemangiomas in Infants and Children. *Surgery*, 71:168-173, 1972.

4. Cunningham, D. S. and Paletta, F. X.: Control of A-V Fistulae in Massive Facial Hemangioma by Muscle Emboli. *J. Plast. Reconstruct. Surg.*, 46:305, 1970.
5. Edgerton, M. T. and Tuerk, D. B.: Macrodactyly (Digital Giantism): Its Nature and Treatment. Symposium on Reconstructive Hand Surgery (Littler, J. W., Cramer, L. M. and Smith, J. W., editors). Vol. 9., St. Louis. The C. V. Mosby Co., Chapter 15:157-172, 1974.
6. Fost, N. C. and Esterly, N. B.: Successful Treatment of Juvenile Hemangiomas with Prednisone. *J. Pediatr.*, 72:351-357, 1968.
7. Frey, H. and Norman, N.: Duration of Action of Depot-corticosteroids. *Pharmacol. Clin.*, 2:109, 1970.
8. Gray, G. R., Freedman, S. I. and Kagan, A. R.: Fibrosarcoma: A Complication of Interstitial Radiation Therapy for a Benign Hemangioma Occurring After 18 Years. *Br. J. Radiol.*, 47:60-61, 1974.
9. Hebley, B. F., et al.: Use of Triamcinolone Acetamide Injections in the Treatment of Allergic Problems. *Ann. Allergy*, 22:244, 1964.
10. Jona, J. Z., et al.: Disseminated Intravascular Coagulation After Excision of Giant Hemangioma. *Am. J. Surg.* 127(5):588-592, 1974.
11. Kwaan, H. C.: Disseminated Intravascular Coagulation. *Med. Clin. North Am.*, 56:177, 1972.
12. Lewis, T. R., Jr.: The Treatment of Hemangiomas. *J. Plast. Reconstruct. Surg.*, 19:201-212, 1957.
13. Li, F. P., Cassady, J. R. and Barnett, E.: Cancer Mortality Following Irradiation in Infancy for Hemangioma. *Radiology*, 113:177-178, 1974.
14. Longacre, J. J., Benton, C. and Unterthiner, R. A.: Treatment of Facial Hemangioma by Intravascular Embolization with Silicone Spheres. Case Report. *J. Plast. Reconstruct. Surg.*, 50:618-621, 1972.
15. Luessenhop, A. J.: Artificial Embolization for Cerebral Arteriovenous Malformations. *Prog. Neurosurg.*, 3:320-362, 1969.
16. Matthews, D. N.: Hemangiomas. *J. Plast. Reconstruct. Surg.*, 41:528-535, 1968.
17. Morgan, J. F.: Use of Sodium Morrhuate in the Management of Hemangiomas. *J. Oral Surg.*, 32:363-366, 1974.
18. Overcash, K. E. and Putney, F.: Subglottic Hemangioma of the Larynx Treated with Steroid Therapy. *Laryngoscope*, 83: 679-682, 1973.
19. Payne, M. M., Moyer, F., Marcks, K. and Trevaskis, A. E.: The Precursor to the Hemangioma. *J. Plast. Reconstruct. Surg.* 38:64, 1966.
20. Prensky, A. L. and Gado, M.: Angiographic Resolution of a Neonatal Intra-Cranial Cavernous Hemangioma Coincident with Steroid Therapy. *J. Neurosurg.*, 39:99-103, July 1973.
21. Weber, G.: The Treatment of Cavernous Hemangioma with Topical Betamethasone 17-Valerate. *Br. J. Dermatol.*, 89:649-651, 1973.
22. Wyman, L. C., Fulton, G. P. and Shulman, M. H.: Direct Observations on the Circulation in the Hamster Cheek Pouch in Adrenal Insufficiency and Experimental Hypercortisolism. *Ann. New York Acad. Sci.* 56:643, 1953.
23. Zarem, H. A. and Edgerton, M. T.: Induced Resolution of Cavernous Hemangiomas Following Prednisone Therapy. *J. Plast. Reconstruct. Surg.*, 39:76-83, 1967.
24. Zweifach, B. W., Shorr, E. and Black, M.: The Influence of the Adrenal Cortex on Behavior of Terminal Vascular Bed. *Ann. New York Acad. Sci.*, 56:626-633, 1953.

DISCUSSION

DR. STEPHEN R. LEWIS (Galveston): Dr. Edgerton does not like Covermark for covering the port wine stain. I'm not sure that his skin grafts are that much prettier than a port wine stain covered with good cosmetics. We have found that the adult male, or most males, will not use Covermark, but females use it, and use it relatively well.

Our skin grafts usually are not as mobile as that skin that he has removed, and I think our end result with cosmetics is usually a little bit better than our skin grafts, although we have skin-grafted a number.

So many times when we have excised the port wine stain, we see a recurrence of the lesion at the border when we're well around the primary lesion. I wonder if Dr. Edgerton has had this same complication or problem with his grafts.

Staged excision in many hemangiomas, with local rotational flaps, if possible, at least on the face, are in our hands a little bit easier and better to use than skin grafts, although a skin graft from the neck does work fairly well.

We have had problems with a number of these cases we have followed over the years that we thought we resolved, and then get rather rapid growth in the patients' thirties or forties especially during pregnancy. I'd like to ask whether steroids in the adult group have been tried by his team, and if they have had any good results. They have not worked well for us.

Lastly, we think very strongly that by using the new method of liquid nitrogen, with probes, in freezing of most hemangiomas of the cavernous type, we have developed good results in a high percentage. Usually, we ligate the external carotids in those intraoral lesions, and then follow it with the nitrogen probe and freezing; and I think this may give us a much better result.

I agree wholeheartedly with his statements on radiation. I think that radiation has caused much more damage than the hemangioma of almost any kind. In one case we had a rather disastrous experience. After ligating the external carotid about 20 years ago on one side, a year ago we ligated the other one, when she had developed massive recurrence in her tongue, buccal mucosa and the cheek, with bleeding, so that we ligated the other external carotid. She then lost about half of her skin over the occiput down to and including part of the bone. Why, we don't know. There is no known reason for the tissue loss, but it's caused us a good deal of trouble.

I do want to congratulate Dr. Edgerton on excellent results with a tremendous number of cases. I would caution that a large percentage of the strawberry marks, if we let them alone and watch them, as Lister said years ago, 95% will disappear without the need of surgery. I do think there is a reason for this disappearance, and we should wait for it in a higher percentage of our cases.

DR. J. ALEX HALLER, JR. (Baltimore): I would like to call attention to a complication of cavernous hemangiomas in children which we have seen in the last several years which Dr. Edgerton did not include in his discussion, and perhaps will mention in closing: This is the problem of platelet trapping in some of these large hemangiomas in children. This process results in thrombocytopenia, with major purpura and even excessive hemorrhage into those tissues surrounding the lesion, as well as from other parts of the body.

This is a particularly terrifying problem. I want to emphasize it this evening because the histologic findings in the biopsy of some of these lesions may be quite misleading. The pathologist may give you the frozen section report that this represents a hemangioendothelioma which is malignant in nature; in other words, a sarcoma of these tissues.

In children a malignant hemangioendothelioma is practically never found. These represent, instead, very active cellular proliferation of the hemangioma, and this activity apparently is related to potential trapping, or adherence, of platelets within the interstices of these vascular masses. They can grow very rapidly. I would like to underline what Dr. Edgerton said about early use of steroids, because we have had some dramatic results in controlling the growth potential of these lesions in children with early, massive steroid therapy.

Two particular cases that we have treated recently were dramatic examples; one, a very large cavernous hemangioma of the buttock of a four-month-old child, would have required practically a hemipelvectomy, and the other in the knee of a little girl of three years of age, which would have required a hip disarticulation. These radical procedures were both recommended by the initial surgeons who saw these children.

Fortunately, they were subsequently biopsied and treated with steroids; both have regressed and, I think, will ultimately disappear. I wanted to emphasize this as one of the rarer but serious complications associated with large cavernous hemangiomas.

DR. RICHARD C. MILLER (Jackson, Mississippi): I rise only to mention some of our experience with visceral hemangiomas, which may be every bit as troublesome as the massive cutaneous and limb hemangiomas which Dr. Edgerton mentioned.

These patients may present with an abdominal mass, (slide) or sometimes they present with hemorrhage. There is little problem, as with this child, when the hemangioma is on the edge of the liver, and it can be resected easily. However, sometimes they are central in the liver, or on occasion they may present with massive hemorrhage. (slide) This child had rectal bleeding from colon involvement; had a pelvis full of hemangioma; and also presented with gross hematuria. He required exploration of his bladder and overseeing of these large hemangiomas inside the bladder. (slide) You are now looking into the bladder, and can see the ulcerated hemangiomas. Eventually the child required rather radical treatment—namely, a total cystectomy. (slide) The pathologic specimen illustrates that the entire bladder is completely infiltrated by hemangioma.

Visceral hemangiomas may or may not regress. Occasionally a child is seen with liver calcifications and we presume he did have a hemangioma. On the other hand, visceral hemangiomas may progress, and require quite vigorous management.

DR. CLAUDE C. COLEMAN, JR. (Richmond): I congratulate Dr. Edgerton on his masterful presentation, but take exception to including a condition as highly malignant clinically, as highly progressive and destructive, as poorly defined roentgenologically, or using any other means of diagnostic acumen, as the congenital arteriovenous malformation.

Our studies, which have been considerable, indicate that about 40% of these tumors are bilateral. We have never been able to delineate the magnitude of the three-dimensional extension of such tumors roentgenologically, because of the vestigial rests, which don't open up until one starts ligating vessels. We feel that this vascular anomaly belongs in a class by itself. Otherwise, I agree with most of what Dr. Edgerton said.

DR. MILTON T. EDGERTON (Closing discussion): Dr. Haller, in the manuscript we do have a considerable section devoted to the problem of the cavernous hemangioma with thrombocytopenia and the development of disseminated intravascular coagulopathy. It's a special and frightening problem.

We have currently one patient now in the hospital that my colleague, Dr. William Futrell, has been struggling to help. This is a young woman, 19 years old. Dr. Claude Coleman saw her at an earlier point in his career when she was only three years old. At our hospital she has just gone through an operation requiring 184 units of whole blood transfusions in just over 72 hours! At present it would appear that she's going to recover. Her disease consisted of a massive cavernous hemangioma of the chest wall, abdominal wall, groin, and lower extremity which was bleeding continuously as a result of D.I.C. with fibrinogen and platelets trapped in the giant angioma.

Paradoxically, heparin was the medication that helped bring the preoperative fibrinogen levels back toward normal in this patient making surgery possible. There's more about D.I.C. in the complete manuscript.

I was delighted to hear that Dr. Haller has also had good results with steroids in treating massive juvenile hemangiomas.

Dr. Lewis correctly makes the observation that most plastic surgeons still recommend some form of makeup to cover many port wine stains. I would make two points to underline my personal disenchantment with these products. Like Dr. Lewis, we have had absolutely *no* success in getting male patients to use make-up of any kind. Don't waste your time! They won't wear it. It shouldn't even be suggested.

It is true that with encouragement some women will use makeup (especially if you don't offer them any surgical alternative). Most of them don't like it, and years later many of them will again seek surgery. They all tell us the same thing: "Doctor, this makeup looks okay, but it takes constant effort, it's hot, and it does nothing to relieve the sense of deformity."

I agree with Dr. Lewis that skin graft replacement of port wine stains is sometimes not a bit better looking than the result with Covermark; but, even so, many patients state they would rather have the skin graft. One stated, "The scars from the skin grafts are the

surgeon's fault. They are not deformities I was born with so I am not embarrassed."

Sometimes the apparently normal skin at the margins of the port wine stains will turn red and form a new rim of capillary hemangioma when you resect all of the clinically involved skin and replace it with a graft. A new area of port wine stain seems to appear in a previously uninvolved area of the skin. Dr. Ferris Smith and Dr. Staige Davis pointed this out many years ago. The only clue we have to the cause of this phenomenon is that it seems to occur in areas of skin that experience maximum tension when the wound is closed after excision of all or part of an angioma. This additional tension may open up vascular filling in the dermis of an occult area (within the predetermined pattern of the angioma) of the vascular anomaly.

We have tried steroids for treatment of *adult* hemangiomas. We have tried them on all age patients with lymphangiomas; and we have tried them to control A/V fistulae. We have had *no* therapeutic success with any of these groups.

I have had limited experience with the liquid nitrogen probe. We have used it only a few times. The results showed skin with uneven

scarring and lumpiness. If the technique can be developed to give uniform thrombosis, it may yet prove of value.

Pregnancy does make some angiomas progress, particularly congenital A/V fistulae. They enlarge markedly, and frightening hemorrhage may occur during the later months of pregnancy.

Many times we should wait for resolution of strawberry marks, but my principal plea tonight is that *each hemangioma* must be individualized. Sometimes surgical excision is simple, quick, effective, and may be the ideal treatment for the child.

Dr. Miller added some valuable comments about visceral hemangiomas which I appreciate, and his experience with these lesions will aid all of us.

Finally, Dr. Coleman, I agree that the A/V fistula embodies *all* of the bad things you said about it. In my opinion the A/V fistula is worse than cancer. I don't mean to suggest that it's like all other hemangiomas even though the pathologist will call it an "angioma" on reviewing the slides. My plea tonight is simply that we must distinguish the A/V fistula from all other varieties of hemangioma if we do call it a type of hemangioma.