The Neuro Image Quiz, Answers

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These images demonstrate a well-circumscribed lobulated erosion of the anterior parietal bone, situated just posterior to the coronal suture. In this case the *zone of transition* from obviously normal bone to obviously pathological bone is very narrow. When the zone of transition is narrower than 3-5 mm on all borders, the lesion usually has low aggressivity. It is expanding slowly, so it is much more likely to be benign. Malignant lesions tend to be permeative and to infiltrate the adjacent bone, producing a wide zone of transition. The answer to question 1 is benign. Analysis of the zone of transition is the basic step in radiological diagnosis of calvarial lesions. There are pitfalls, however. First, a malignant lesion may exhibit a narrow zone of transition on nearly all sides when it develops as malignant degeneration of a previously benign lesion, e.g. sarcomatous degeneration of fibrous dysplasia. Second, a benign inflammatory/infective lesion may permeate widely producing a very wide zone of transition. Thus, infection can be very difficult to differentiate from malignancy. Nonetheless, analysis of the zone of transition usually distinguishes benign from malignant lesions [1].

Within the group of benign lesions, the speed of lesion growth can be assessed by reviewing the degree of sclerosis along the margin of the lesion. Very slowly growing lesions afford time for the calvarium to enclose the lesion in a dense rim or ‘rind’ of sclerosis. Dermoid and epidermoid tumors commonly exhibit such a rind. So do some hemangiomas. Benign but more aggressive lesions expand more rapidly and afford less time for deposition of bone, creating a narrow (pencil-thin) rim of bone. More active, but still benign, lesions exhibit a sharp zone of transition with no sclerotic rim at all. Eosinophilic granuloma is an aggressive benign lesion and typically shows a sharp zone of transition with little or no rim of reactive sclerosis.

When lesions erode the inner and outer tables of the calvarium to a different degree, the margins of the calvarial defect are said to be beveled. Lesions arising external to the calvarium typically erode the outer table more than the inner table and are ‘beveled-from-without’. Such lesions include anterior fontanelle dermoids, some stages of cephalhematoma deformans and eosinophilic granuloma. Other lesions erode the inner table more

than the outer table and are said to be beveled-from-within. Such lesions include calvarial erosion by arachnoid cyst, intracranial glioma and some encephaloceles.

In the patient illustrated here, the narrow zone of transition, very thin reactive cortical sclerosis and beveled margins suggest that the lesion is most likely an eosinophilic granuloma. The answer to question 2 is F, eosinophilic granuloma.

Surgery disclosed an encapsulated mass situated deep to the galea and separated from the galea by loose alveolar tissue. The capsule was yellowish to dark brown and resembled inflamed granulation tissue. The lesion core was chocolate, necrotic tissue. The deep layer of the capsule was attached to the bone. Sparse finger-like extensions of the lesion into bone required focal craniectomy. The deepest portion of the mass had eroded through the inner table to dura, forming a very thin epidural mass that did not undermine the surrounding normal bone. Histologic examination revealed fibroconnective tissue infiltrated with masses of histiocytes, giant cells, foam cells and hemosiderin-laden cells, plus areas of hemorrhage and necrosis. Immunohistochemical stain for S100 was positive. Final pathological diagnosis was Langerhans' cell histiocytosis. This term is now the preferred name for the histiocytosis traditionally called eosinophilic granuloma.

Rawlings and Wilkins [2] studied 26 cases of solitary eosinophilic granuloma of the skull. All the patients had solitary enlarging tender scalp masses. The parietal bone was affected most commonly, but any bone could be involved.

In our experience, about 40% of eosinophilic granulomas extend to the dura. They may undermine slightly between the inner table and the dura forming a slight epidural mass. The eosinophilic granuloma usually remains epidural, but may invade through the dura into the cerebral cortex causing Jacksonian seizures in exceptional cases [3]. Approximately one third of patients will develop new lesions during follow-up. Younger patients have a higher risk of recurrence.

Analysis of the experience at Children's Memorial Hospital, Chicago, and review of the literature indicates that most nontraumatic lumps of the head are benign lesions. Choux et al. [4] found that 33 of 36 calvarial lesions

sions (92%) were benign. At our hospital, 70 consecutive nontraumatic lumps of the head have proven to be benign [5]. The answer to question 3 is true. The commonest benign lump of the calvarium is an (epi)dermoid tumor. The answer to question 4 is E, der-moid [4,5]. (Epi)dermoids constituted 31% of the series of Choux et al. [4] and 61% of our personal material. Previously unsuspected cephalhematomas deformans constituted 9% of our material, eosinophilic granuloma 7%, and occult meningoencephaloceles 4%. Hamartomas, hemangiomas and fibrous dysplasia each accounted for 3% of nontraumatic head lumps. The remaining lesions were truly miscellaneous – a single instance each of a wide spectrum of pathology [5]. True benign tumors of the calvarium are rare. The commonest such tumor is the osteoma [6]. We observed no case of a primary benign tumor in 70 patients with head lumps, perhaps because calvarial tumors are more frequent in older children and adults. Primary malignant tumors of the calvarium are even rarer than the primary benign tumors [4, 6]. The most frequent primary malignant tumor is the osteogenic sarcoma. In our experience, approximately 61% of nontraumatic head lumps are related to a cranial suture. The suture involved is most commonly the frontozygomatic suture. The lesion most commonly found along a suture is the (epi)dermoid. In our personal series, 86% of (epi)dermoids lay along a suture; 86% of perisutural lesions were (epi)dermoids. Eosinophilic granulomas, hemangiomas, hamartomas, nasal gliomas and occult meningoencephaloceles were also found to lie along sutures. In the patient illustrated here, the contiguity of the lesion with the coronal suture was a misleading coincidence. Our case material shows that the edge of an eosinophilic granuloma reaches to or overlies a cranial suture in 2 of 6 cases. Therefore, perhaps one third of eosinophilic granulomas may have a (presumably) coincidental relationship to a suture. Significant intracranial extension of a calvarial lesion can be defined as (a) an intracranial component that exerts mass (whether extradural or intradural) and (b) extension intradurally (whether it exerts mass or not, e.g.

dermal sinus). In our personal experience with 70 cases, 63% of head ‘lumps’ remained in or external to the calvarium. Twenty percent of lesions reached the outer layer of dura but exerted no substantial intracranial mass effect. These lesions were judged to have no significant intracranial extension. However, 1% of head lumps were the external sign of a major extradural mass and 16% of head lumps had definite intradural extension, with or without mass. Thus, 17% of children with nontraumatic head lumps proved to have significant intracranial extension. The answer to question 6 is B, 15%. Histologically, the lumps associated with significant intracranial extension were scalp/epidural abscesses (1), dermoid tumors (5), occult meningoencephaloceles (3), hamartomas (2) and calvarial fenestration by tumor-associated hydrocephalus (1). In this last case, maternal detection of scalp-bulging over a second ‘soft spot’ led to detection of a choroid plexus papilloma, extreme hydrocephalus, and protrusion of the cerebral mantle through an eroded calvarium.

References

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