Subcutaneous neuroglial choristoma: an immunohistopathological case study

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Congenital nests of neuroepithelial tissue, probably representing either sequestered encephalocele or aberrant neuroectodermal migration in utero, may give rise to heterotopic neuroglial tissue masses.¹ The 'nasal glioma' is best known, although small subcutaneous heterotopic neuroglial masses may occur near the midline on the head and face,¹² or in other locations, such as the chest wall.³ Periocular heterotopic neuroglial tissue has been documented in the orbit,^{4,5} in a lower eyelid,⁷ and with limbal dermoid.^{8,9} There may be associated central nervous system malformations.⁷ Neuroglial choristomas are heterotopic neuroglial masses with no clinical,

radiological, or histopathological evidence for origin from sequestered encephalocele.

Case report

A 3-month-old male infant had had a subcutaneous mass at the temporal end of the left eyebrow since birth. This measured 1.5 cm on examination. Dermoid cyst was diagnosed, and excision was attempted. At surgery, the lesion was more fleshy than a typical dermoid cyst. A subtotal excision was performed. Postoperative computed tomography showed no connection to the intracranial contents (Fig 1). The eyes, optic nerves, and brain were normal.

Seven weeks later the remaining lesion was excised. By then the residual supraorbital mass had grown, and a similar lesion was seen near the centre of the patient's forehead. This second lesion measured 1.5 cm; it was not contiguous with the first. Gross total excision of both lesions was achieved at the second surgery. Wide excision was not performed because of the possible reconstructive surgery that such a procedure might necessitate.

The three surgical specimens were formalin fixed and paraffin embedded. Sections were studied with haematoxylin and eosin stain and the peroxidase antiperoxidase method for S100 protein and glial fibrillary acidic protein (GFAP).¹⁰ The immunohistochemical chromogen was diaminobenzidine, which produces a dark brown stain.

All specimens showed glial cell infiltration of subcutaneous fibrous tissue and skeletal muscle (Fig 2). The intermingled glial cell processes resembled neuropil. These cells had round to ellipsoid nuclei with finely divided chromatin, occasional small nucleoli and smooth nuclear membranes. The pale eosinophilic cytoplasm was finely fibrillated. The glial cytoplasm and nuclei were S100 protein positive (Fig 3A), and cytoplasmic filaments were GFAP positive (Fig 3B). There were no meningotheelial cells, ependymal cells, or teratomatous elements; a very rare cell suggestive of a neuron was seen.

There was no further treatment. After 3 months, both lesions recurred locally. They grew to about the same size as before, and then remained stable over a 2 year follow up period. The child has been otherwise healthy and normal.

Comment

Subcutaneous heterotopic neuroglial tissue occurs most often in male children younger than 2 years of age, presenting as a painless, static, or slowly growing lesion.¹ Dermoid cyst is a frequent preoperative diagnosis. The differential

Figure 1 Unenhanced computed tomographic (CT) scan after the first surgical procedure, showing subcutaneous lesion over left superior orbital rim. Note that the underlying bone is intact.

Figure 2 Benign glial cells with round to ellipsoidal nuclei and fibrillated cytoplasm infiltrate the fibrous subcutaneous connective tissue in this field. (Haematoxylin and eosin, magnification $\times 70$.)

Fig 3A

Fig 3B

Figure 3 In these fields the glial tissue is highlighted by nuclear and cytoplasmic staining for (A) S100 protein (peroxidase antiperoxidase with diaminobenzidine, magnification $\times 70$), and by staining of the glial cytoplasmic filaments for (B) GFAP (peroxidase antiperoxidase with diaminobenzidine, magnification $\times 70$). The positively stained areas are dark brown.

diagnosis includes haemangioma, fibroma or fibromatosis, and lipoma.

All forms of heterotopic neuroglial tissue are histologically similar, and contain mostly glial tissue, although neurons, ependymal cells, or meningotheelial cells are occasionally seen.¹² The neuroglial tissue tends to be locally infiltrative.

Old lesions in adults may be fibrotic.¹² Several previous articles describe positive immunohistochemistry for S100 protein or GFAP in heterotopic neuroglial tissue.^{2,3,7,11,12} These methods provide sharp contrast between neuroglial tissue and connective tissue (Figs 3A, B), and are useful in fibrotic cases.²

Complete excision is the suggested treatment for neuroglial choristoma. Differentiating heterotopic neuroglial tissue (choristoma or sequestered encephalocele) from encephalocele with intracranial communication solely on histopathological or immunohistochemical grounds may be impossible, so clinical and radiological evaluation is important.^{1,13,14} Coordination with a neurosurgical consultant is often necessary when a communicating encephalocele is suspected. When biopsy reveals a benign subcutaneous neuroglial lesion, with clinical and radiological features of neuroglial choristoma, our experience suggests that close clinical follow up alone may also be a reasonable approach.

The opinions and assertions contained herein are the private views of the authors and should not be construed as being official or as representing the views of the Department of the Army or the Department of Defense.

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