Cranial fasciitis: presentation as a post-auricular mass

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Abstract

Cranial fasciitis is a rare variant of nodular fasciitis. It is a benign condition with features resembling sarcoma, seen principally in young males. It involves the skull bones and grows at a rapid pace. Accurate diagnosis and surgical excision is the key to management. Prognosis is good with recurrence rare. By 1992, 17 cases had been reported in the literature. The present case is the first reported in Pakistan. It presented as a post-auricular mass.

Key words: Fasciitis, cranial; Scalp

Case report

A five-year-old girl was admitted, to the ENT Department at the Civil Hospital, Karachi, with a right post-auricular swelling of five months duration. She had otorrhoea and intermittent pain in the right ear. No history of trauma was reported. Examination showed a soft swelling 3×2.5 cm behind the right ear with poorly delineated margins. The swelling was non-fluctuant and lacked signs of acute inflammation. A small central perforation of the tympanic membrane was noted with margins showing circumscribed congestion and a scanty mucopurulent discharge. A partial right infra-nuclear facial palsy was also present.

Routine investigations were all normal save for a probable bony erosion observed on plain mastoid films. Wide bore needle biopsy did not yield any aspirate and the patient was admitted for surgery with a provisional diagnosis of post-auricular abscess. On exposure through a post-auricular incision a large dehiscence was found in the temporal bone with an intact lateral sinus bulging through it. More extensive surgery was deferred pending further investigation. CT scan showed a space-occupying lesion in the posterior and middle cranial fossa with erosion of the petrous temporal bone (Figure 1).

A right carotid angiogram showed increased vascularity in the temporal bone region and the venous phase showed occlusion of the distal portion of right transverse sinus (Figure 2). While these investigations were underway a soft pinkish mass protruded through the post-auricular incision (Figure 3). This was biopsied by the neurosurgeon and a tissue diagnosis of cranial fasciitis was established. Definitive surgery was planned. On exposure soft, relatively avascular, extradural tissue was found in the posterior and middle cranial fossa involving petrous and mastoid parts of the temporal bone. Complete excision of the mass was attempted with currettage of the involved bone. No evidence of inflammatory disease was found in the mastoid air cells but a small collection of pus was noted in the antrum. Recovery was uneventful and the patient was discharged eight days later.

Histology revealed proliferation of spindle and skeletal fibroblast cells set in a myxoid matrix. In some places a starriform pattern was seen along with some areas of fibrosis and acute inflammation (Figures 4 and 5). Skeletal muscle fibres and calcification were also seen but there was no evidence of malignancy. These features are compatible with cranial fasciitis. Regular follow-up confirmed complete recovery of the facial palsy after one month and no recurrence at six months.

Discussion

Cranial fasciitis is a variant of nodular fasciitis (Kyriakos, 1990), which is a proliferative fibroblastic lesion presenting as a tumour-like mass (Davies *et al.*, 1989). Nodular fasciitis is mostly seen in the extremities, trunk, face and neck (Frei *et al.*, 1991). It develops from the subcutaneous muscle tissue and histologically it mimics sarcoma.

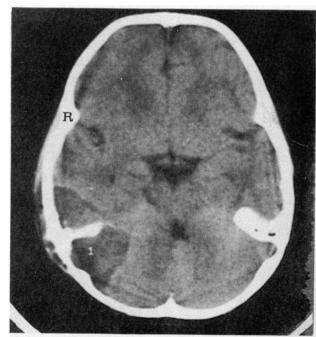
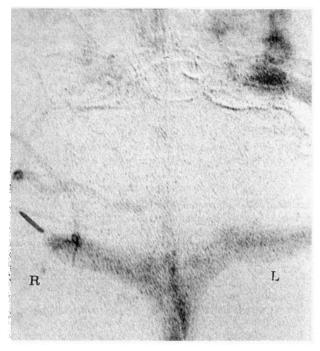


Fig. 1

CT scan showing space-occupying lesion in the posterior and middle cranial fossa with erosion of the petrous temporal bone. (R: right-hand side).

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 $${\rm Fig.}~2$$ Venous phase of the carotid angiogram showing occlusion of the distal portion of right transverse sinus. (R: right-hand; L: left-hand side).



 $$\operatorname{\textsc{Fig.}}$3$$ Cranial fasciitis emerging as a post-auricular mass.

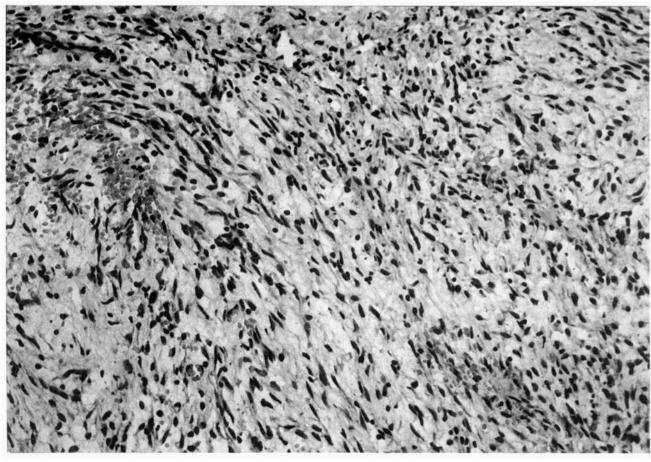


Fig. 4

Fibroblasts arranged in short interlacing bundles and fascicles. Compact areas of cellularity well marked. Fibroblastic cells intermingled with inflammatory cells and extravasated red blood cells. (H&E; ×100).

CLINICAL RECORDS 257

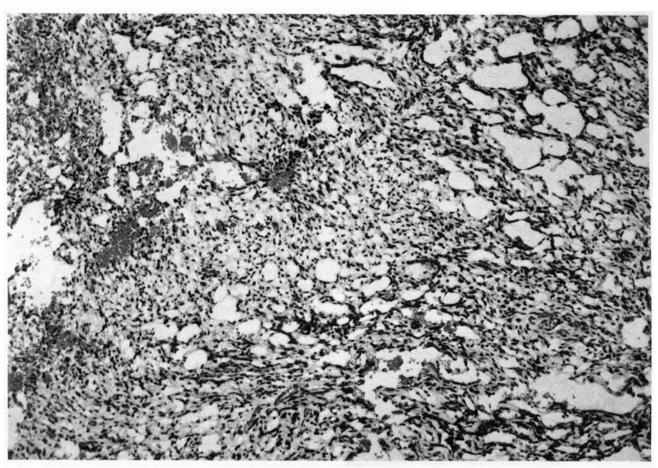


FIG. 5
Cellular areas showing mucoid pools separating fibroblastic cells. (H&E; ×100).

Cranial fasciitis is a benign lesion differing from the typical nodular form by virtue of its proximity to the skull. Uncertainty prevails about the exact site of origin of cranial fasciitis. Lauer and Enzinger (1980) mention that origin from one of the deep fascial layers of the scalp, or the underlying periosteum seems most likely. It shows a male predilection and exhibits the tendency to grow rapidly and painlessly in the scalp. In most cases it destroys the outer tables of the skull, and has been found adherent to the dura (Koyama et al., 1991). A possible aetiological factor is considered to be a reactive phenomenon secondary to trauma (Schiller, 1988). The lesion has a benign clinical course and surgical excision with, or without, currettage of the underlying bone definitely cures it (Patterson et al., 1989). Recurrence is uncommon. Histologically it has been described as a loose proliferation of stellate to spindle-shaped fibroblasts in a myxoid background with foci of haemorrhage and hyalinization (Coates et al., 1990). Kumon et al. (1992) reported cranial fasciitis in the left frontal bone invading the anterior skull base. Cranial fasciitis involving the occipital bone was reported by Mollejo et al. (1990) and Inamura et al. (1991). Koyama et al. (1991) reported cranial fasciitis in a one-year-old boy arising in the temporoparietal region.

By 1992 17 cases of cranial fasciitis had been reported in the literature (Kumon *et al.*, 1992). We report an additional case of this rare disease with involvement of the temporal bone, presenting as a post-auricular mass. As far as we know this case is the first in Pakistan.

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