Intracranial fibro-osseous lesion

Case report

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A rare tumor closely associated with the trigeminal ganglion produced a syndrome of atypical facial pain in a 44-year-old man. Two previous reports have designated similar tumors as "fibro-osseous lesions." The distinctive morphological appearance is illustrated and possible histogenesis discussed.

KEY WORDS • fibro-osseous lesion • intracranial neoplasm • trigeminal nerve • facial pain

FIBRO-Osseous lesions of the intracranial space are rare. Two previous reports have described seven possible cases of this disorder. Distinct from the ossifying and nonossifying fibromas of bone, they have a characteristic morphological appearance. This report details the case of a 44-year-old man presenting with a tumor closely associated with and, at least peripherally, involving the trigeminal ganglion. The appearance of this unusual tumor is illustrated and the immunohistochemical findings are discussed.

Case Report

This 44-year-old man had a 6-year history of atypical right facial pain. The pain involved the face in all three divisions of the trigeminal nerve and included the right ear and the temporomandibular and occipital areas. No specific precipitating factors or trigger areas were noted by the patient. Pain was described as stinging and constant with intermittent acute exacerbations. Marcaine (bupivacaine) occipital nerve blocks and medication were only partially effective. Two temporomandibular joint arthroplasties were performed with removal of small calcified osteophytes, but pain relief was only temporary. Transection of the greater occipital nerve on the right and subsequent intradural right-sided C-2 and upper C-3 dorsal rhizotomy in another hospital resulted in resolution of the occipital pain but the ear and facial pain remained. A computerized tomography (CT) scan showed a 1-cm calcified mass impinging on the right trigeminal ganglion (Fig. 1).

Examination. The patient's admission examination was unremarkable except for a mild decreased response to pinprick in the right mandibular division of the fifth cranial nerve. No history of trauma was elicited.

Operation. On exploration of the trigeminal ganglion region, a 1-cm mass composed of both firm and soft components was encountered. There was no clear-cut connection to bone, but the mass was adherent to the dura and to the adjacent nerve. The mandibular division of the fifth nerve was sacrificed in order to completely remove the tumor. After the operation complete relief of pain was reported by the patient.

Pathological Findings. The portion of tumor received for frozen section was examined and a diagnosis

Fig. 1. Computerized tomography scans, coronal view (a) and axial view (b), showing a high-density lesion in the region of the right trigeminal ganglion. Although the lesion was apparently in contact with bone, surgical exploration demonstrated no definite attachment.
of probable meningioma given. Additional fragmented portions of the firm tumor were received for preparation of permanent sections.

Paraffin-embedded sections were stained with hematoxylin and eosin, Alcian blue, Masson's trichrome, periodic acid-Schiff (PAS), von Kossa, alizarin red, Verhoeff-van Gieson, and Wilder's reticulin. Immunostains for S-100 protein, glial fibrillary acidic protein (GFAP), vimentin, laminin, α1-antitrypsin, lysozyme, keratin, and cytokeratin were made using commercial antibodies and a Vectastain avidin-biotin-peroxidase complex (ABC) kit.

The tumor had a connective tissue background enclosing multiple variable-sized irregular foci of amorphous gray-blue material. The material in these foci appeared as multiple rounded or angulated plates of varying size. Spindle cells and occasional bi- or multinucleated giant cells were noted at the edges of the plates (Fig. 2a). Focal areas of intense hematoxylin staining were present in the amorphous material. The material was rimmed by a palisade of spindle cells with elongated hyperchromatic nuclei. Occasional multinucleated cells were also present. The fibrous tissue separating the amorphous areas contained similar spindle cells, some mildly pleomorphic (Fig. 2a inset). No mitoses were noted. Trabecular bone formation was present in the periphery of some fragments (Fig. 2b). Reticulin stains demonstrated a diffusely positive background with concentration and fragmentation within the collection of amorphous material, which also stained with Alcian blue, PAS, von Kossa, alizarin red, and blue with Masson's trichrome stain. Vimentin immunoperoxidase stains were positive in the cytoplasm of the palisading cells (Fig. 2c). Laminin stains were strongly positive in the background spindle cells, weakly positive in the palisading spindle cells, and negative in the amorphous material. Keratin, cytokeratin, S-100 protein, α1-antitrypsin, lysozyme, and GFAP immunostains were negative. Normal ganglion cells from the trigeminal ganglion were seen at the periphery of one fragment.

The irregular plates noted at the light microscopic level appeared to correlate with masses of electron-dense amorphous material at the electron microscopic level (Fig. 3a). Many of the smaller masses had a central accumulation of collagen fibrils (Fig. 3b). Cells at the periphery contained abundant intracytoplasmic filaments or prominent rough endoplasmic reticulum. They had no junctional attachments but had smooth cytoplasmic borders. Spherical nodules of the form noted by Rhodes and Davis 6 were not present, although it is possible that the smaller masses with collagen centers may represent a variation of these nodules. Extracellular basal lamina material was also present (Fig. 3c).

**Discussion**

Only two previous reports have detailed the unique features of intracranial fibro-osseous lesions. 3,6 These lesions have been defined by accumulations of a peculiar amorphous material surrounded by spindle cells in conjunction with a variable amount of bone formation. The majority of lesions were incidental findings at autopsy. However, their capacity to proliferate and induce symptoms is illustrated in Case 1 of Rhodes and Davis, 6 in the case reported by Jun and Burdick, 3 and the present case. The rate of growth is apparently slow. The lesion bears a morphological resemblance to those seen in tumoral calcinosis or secondary hyperparathyroidism. 7 However, those lesions do not contain bone and the material in the central amorphous regions consists of calcium phosphates and carbonates. 4 One electron microscopic study of tumoral calcinosis demonstrated no structures with collagen fibril centers or associated basal lamina material as seen in our study. 3 Unfortunately, serum calcium and phosphate levels were not measured during the hospitalization of our patient. The lack of α1-antitrypsin or lysozyme

![Fig. 2. Photomicrographs of the lesion. a: Spindle cells can be seen palisading about the periphery of a nodule of amorphous material. H & E, × 100. Inset: High-power view of palisading cells. H & E, × 275. b: Bony trabeculae are seen at the periphery of the lesion. H & E, × 150. c: View showing palisading cells with granular cytoplasmic staining. Vimentin, × 200.](image)
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cells with vimentin and laminin supports but does not distinguish between these two possibilities. The suggestion that this lesion represents abortive membranous bone formation is an interesting one. Our electron microscopic examination does show structures within the amorphous material with collagen fibers at their centers and amorphous peripheries that appear similar to those found in early membranous bone.\(^1\) However, this explanation does not fully account for all the features of the tumor. Bone formation is an integral part of the lesion. If the amorphous material represents defective bone formation, it would have to be a focal failure. The amorphous material does not seem to serve as a nidus of bone formation, since no transitions between the material and bony trabeculae were observed. The nature of the palisading cells surrounding the material and the precise function they fulfill in either formation of, or in reaction to, the material also needs further clarification.

Acknowledgment

We would like to acknowledge the assistance of Dr. John Kepes of the University of Kansas in making the original diagnosis.

References


Manuscript received April 22, 1988.
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