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Nasopharyngeal Nonossifying Variant of Ossifying Fibromyxoid Tumor: CT and MR Findings

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Summary: The CT and MR findings of a nasopharyngeal nonossifying variant of an ossifying fibromyxoid tumor are presented. The findings are radiographically indistinguishable from more common malignant neoplasms encountered in this region. The tumor was isointense with muscle on T1-weighted images. On proton density– and T2-weighted images, the mass was mostly isointense with gray matter but contained some areas of lower intensity that might reflect the fibrous tissue component. The tumor eroded through the floor of the middle cranial fossa.

Index term: Nasopharynx, neoplasms

The nonossifying variant of an ossifying fibromyxoid tumor is a very rare variant of the ossifying fibromyxoid tumor. The ossifying variant is well documented and typically occurs in the extremities of adult men (1). The histogenesis is unclear, although a Schwann cell origin has been proposed (2). We describe a case of an aggressive form of a nonossifying variant of an ossifying fibromyxoid tumor involving the base and nasopharyngeal space of the skull in an adult man.

Case Report

A 35-year-old man, previously in good health, presented to an outside institution after 6 months of left-sided nasal congestion and pain referable to the distribution of all three divisions of the left trigeminal nerve. A large nasopharyngeal soft-tissue mass was visualized on physical examination. Palsies of cranial nerves III, IV, and VI on the left were present. At the local hospital, a contrast-enhanced computed tomographic (CT) scan showed a heterogeneous, enhancing mass involving the left parapharyngeal space that projected into the nasopharynx. The initial biopsy was interpreted as neurofibroma. The patient was referred to our hospital, where a repeat transnasal biopsy revealed a spindle cell tumor, thought to represent myofibromatosis. An arteriogram showed the lesion to be hypovascular. Surgical resection was attempted, but because of unexpected bleeding, only a small portion of tumor was removed.

Fifteen months later, a CT scan showed a contrast-enhancing tumor extending into the left middle temporal fossa and temporal lobe. The posterior left ethmoid sinus and sphenoid sinus, as well as the left posterior choana, nasopharynx, and eustachian tube, were filled with tumor (Fig 1A). On magnetic resonance (MR) imaging, the tumor was isointense with muscle on T1-weighted images. On proton density– and T2-weighted images, the mass was mostly isointense with gray matter but contained some areas of lower intensity. Contrast-enhanced MR clearly showed extension of the deeply enhancing mass into the left middle cranial fossa secondary to erosion of its floor and involvement of the left cavernous sinus, presumably because of tumor extension via the foramen of ovale (Fig 1B–1D).

On histologic examination, the tumor contained small, cytologically bland spindle to oval cells and delicate capillaries present in a fibromyxoid matrix surrounded by dense connective tissue. In areas, collagen fibrils separated the homogeneous cells into interweaving fascicles. Mitotic activity and osteoid/bone were absent (Fig 2). A Bodian stain for nerve fibers was negative; therefore, the tumor was not considered to be of neural origin. The absence of bone/osteoid elements and review by the Armed Forces Institute of Pathology confirmed the impression that this tumor was a "nonossifying" variant of an ossifying fibromyxoid tumor.

Radiation therapy was offered, but the patient was unavailable for follow-up.

Discussion

Ossifying fibromyxoid tumors are a rare and only recently described soft-tissue neoplasms. In 1989, Enzinger et al (3) reported that 59 cases were collected by the Armed Forces Institute of Pathology over a 25-year period. The tumors were previously classified under various

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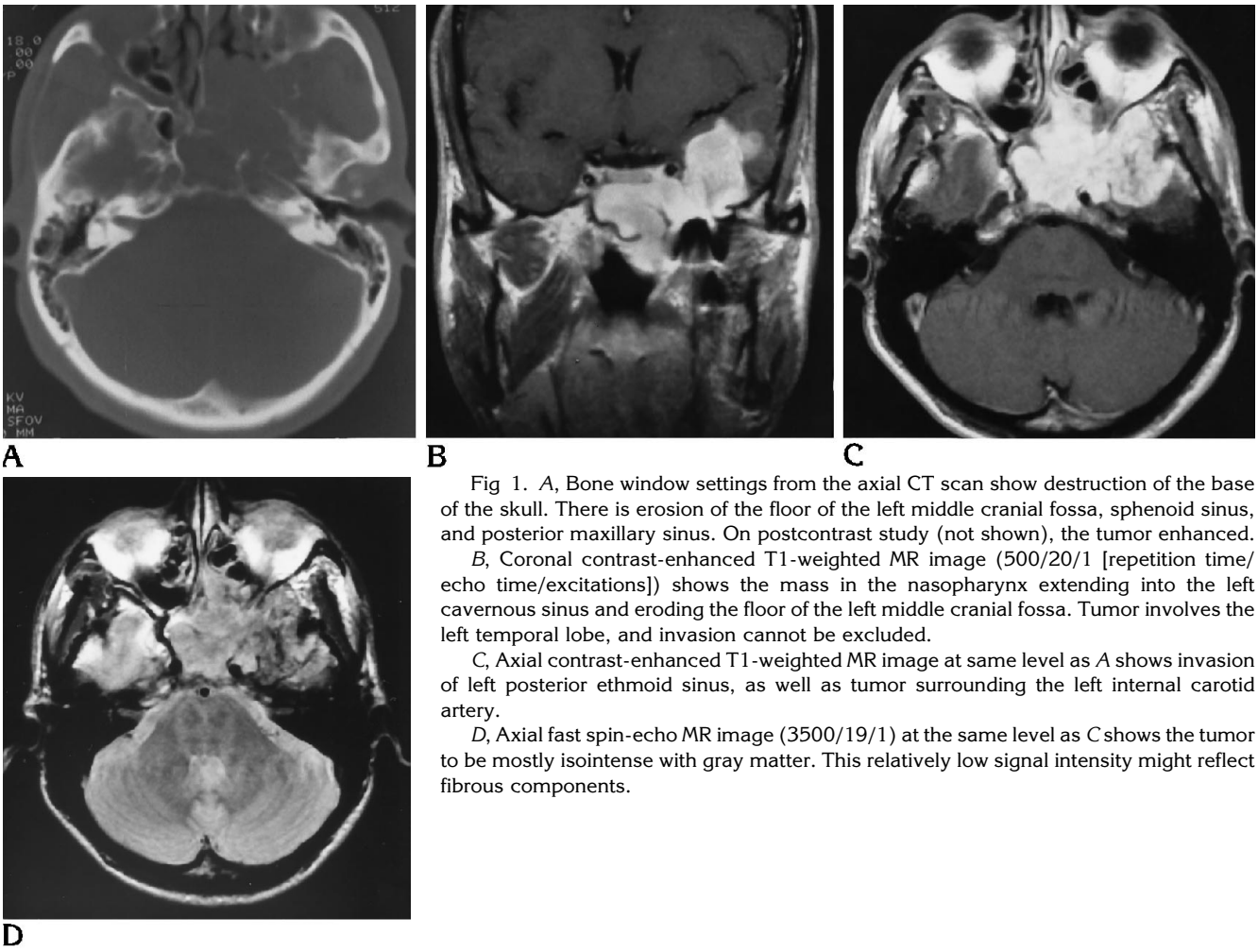

Fig 1. *A*, Bone window settings from the axial CT scan show destruction of the base of the skull. There is erosion of the floor of the left middle cranial fossa, sphenoid sinus, and posterior maxillary sinus. On postcontrast study (not shown), the tumor enhanced.

B, Coronal contrast-enhanced T1-weighted MR image (500/20/1 [repetition time/echo time/excitations]) shows the mass in the nasopharynx extending into the left cavernous sinus and eroding the floor of the left middle cranial fossa. Tumor involves the left temporal lobe, and invasion cannot be excluded.

C, Axial contrast-enhanced T1-weighted MR image at same level as *A* shows invasion of left posterior ethmoid sinus, as well as tumor surrounding the left internal carotid artery.

D, Axial fast spin-echo MR image (3500/19/1) at the same level as *C* shows the tumor to be mostly isointense with gray matter. This relatively low signal intensity might reflect fibrous components.

categories that included tumors of osteoblastic, neural, and chondroblastic origins (3). They occur almost exclusively in adults, predominantly in men. Typically, the lesions are painless and small, and most arise within the deep subcutis or muscle (4). The vast majority involve the upper and lower extremities and less frequently the trunk.

Ossifying fibromyxoid tumors are generally slow-growing masses and are histologically benign or of low-grade malignancy. The tumors are generally well circumscribed and consist of lobulated nests of small, round cells within a myxoid to hyalinized stroma surrounded by dense fibrous tissue. In general, they are highly vascularized. Most exhibit a thin, incomplete shell of lamellar bone in the capsular region (5). All tumors are at least partially circumscribed by a thickened, cartilaginous capsule. Occasionally, a transition toward a low-grade osteosarcoma with abundant osteoid formation is seen in recurrent tumors (3).

The tumor presented here is a rare variant called a nonossifying ossifying fibroma. This variant is devoid of a rim of bone or bone-forming elements and occasionally displays an increased mitotic rate (4). It is difficult to distinguish from myofibromatosis. Surgery is the treatment of choice, but local recurrence can be seen in at least 27% of patients (3).

In 1992, Williams et al (4) reported two cases of the nonossifying variant involving the head and neck. Of these, one was frankly malignant and arose in the left parapharyngeal space in a 58-year-old man. In our patient, despite histologic absence of malignancy, the tumor behaved aggressively. Even with an angiogram that showed the tumor to be hypovascular, excessive bleeding occurred during surgery and prevented complete resection.

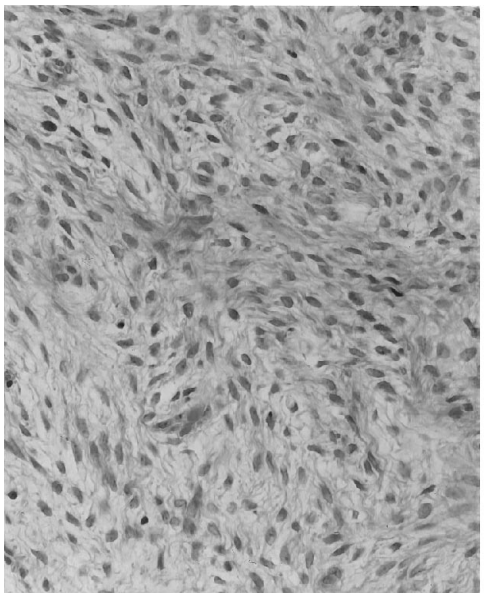

Fig 2. On photomicrograph (hematoxylin-eosin, ×300) of histologic section, the tumor contains small, uniform spindle cells arranged in a swirling pattern embedded in dense connective tissue compatible with a fibromyxoid tumor. No bone was found, suggesting the nonossifying nature of the lesion. Mitoses were infrequent.

In summary, the tumor described here was radiographically indistinguishable from more common malignant neoplasms encountered in this region such as squamous cell carcinoma, adenocarcinoma, adenoid cystic carcinoma, sarcoma, and metastatic tumors. Its low signal intensity on T2-weighted sequences, similar to that seen in some high-grade malignancies of the nasopharynx, probably reflects the fibrous tissue component. The nonossifying variant of ossifying fibroma is a very rare tumor, and biopsy is required to establish a definite diagnosis.

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