Benign fibroosseous lesions involving the skull base, paranasal sinuses, and nasal cavity

Report of two cases

KIYOSHI SAITO, M.D., KEIZO FUKUTA, M.D., MASAKATSU TAKAHASHI, M.D., YUKIO SEKI, M.D., AND JUN YOSHIDA, M.D.

Departments of Neurosurgery and Otorhinolaryngology, Nagoya University School of Medicine, Nagoya, Japan; and Department of Plastic and Reconstructive Surgery, Komaki City Hospital, Komaki, Japan

The authors report two patients with benign fibroosseous lesions involving the center of the skull base: a 15-year-old boy with repetitive meningitis and pneumocephalus and a 11-year-old boy with nasal obstruction and headache. The clinical diagnoses were ossifying fibroma and aneurysmal bone cyst, respectively. Lesions in both patients extended to the nasal cavity, the sphenoid and posterior ethmoid sinuses, and the skull base; where the planum sphenoidale, the sella turcica, the upper two-thirds of the clivus, and the medial portion of the middle cranial base were involved. The lesions were totally removed using an anterior craniofacial approach. Characteristics of these lesions and the surgical approach are discussed.

KEY WORDS • aneurysmal bone cyst • ossifying fibroma • surgical approach • benign fibroosseous lesion • skull base

A benign fibroosseous lesion is a bony lesion in which bone is replaced with benign fibrous tissue containing various proportions of mineralized structures. This term has been used mainly to designate both fibrous dysplasia and ossifying fibroma, because no universally accepted histological criteria categorically distinguish these lesions from one another. It has been stated that ossifying fibroma differs from fibrous dysplasia only in showing a sharper demarcation from adjacent normal tissue. Other lesions in this category include cementifying fibroma, cementoossifying fibroma, benign osteoblastoma, and aneurysmal bone cyst.

Ossifying fibromas are found almost exclusively in the craniofacial bones, with only rare reports of long-bone involvement. Among the craniofacial bones, the mandible is the most common site, accounting for 75 to 89% of cases. Although less commonly, calvaria, orbit, maxilla, or paranasal sinuses have also been involved. On the other hand, aneurysmal bone cysts are usually found in long bones or in the spine and rarely occur in the skull (3–6% of cases) and facial bones. The midline of the skull base is not a site of predilection for either ossifying fibroma or aneurysmal bone cyst. We present two massive benign fibroosseous lesions occurring in the skull base, including an ossifying fibroma and an aneurysmal bone cyst, both of which were totally removed using the anterior craniofacial approach described by Spetzler, et al.

Case Reports

Case 1

History. This 15-year-old boy presented with a 2-month history of repetitive meningitis and progressive deterioration of left visual acuity over several years.

Examination. The patient’s visual acuity was 20/30 on the right and finger counting on the left. Computerized tomography (CT) and magnetic resonance (MR) images revealed marked pneumocephalus and a large multilobulated tumor involving the nasal cavity, sphenoid bone, and skull base (Fig. 1). In the skull base, the planum sphenoidale, sella turcica, upper clivus, and right middle cranial base were involved. The tumor had an outer shell and internal septa of bone density with varying thickness. Thick portions of the shell showed strong enhancement after injection of gadolinium. Because the lesion was sharply demarcated, the clinical diagnosis was ossifying fibroma.

Operation. After effective treatment of meningitis with antibiotic medication, surgery was performed using the anterior craniofacial approach described by Spetzler, et al. Through a bicoronal incision and a bifrontal craniotomy with right temporal extension, frontonasoorbital bone, including both superior orbital rims and orbital roofs, was removed as a unit. After being mobilized by circumferential osteotomy, the cribriform plate was elevated with the olfactory mucosa and the frontal dura. Using an epidural approach, the compressed dura mater was separated from the lesioned bone and was followed from the planum sphenoidale to the tuberculum sellae, sellar floor, dorsum sellae, and the clivus. Both optic canals were released. The optic nerves showed compression and thinning that was worse on the left side. The interomedial dural walls of the cavernous sinus on both sides and the pituitary dura were thoroughly exposed. The tumor was multilobular.
Anteriorly, cysts had walls of fibrous tissue or mucosal membranes and contained xanthochromic fluid. Posteriorly, the solid portion was brown, granular, and relatively vascular. Total removal of the tumor was achieved. A dural laceration was found medial to the left optic nerve and was sutured.

The skull base was reconstructed using a 10-cm-wide bipedicled temporoparietal galeal flap based on the superficial temporal arteries. The flap was wide enough to cover the entire defect of the skull base. The cribriform plate, the frontonasoorbital unit, and the frontotemporal bone were repositioned and affixed using wires and mini- and microplates.

**Postoperative Course.** The postoperative course was uneventful except that the patient experienced decreased olfactory function, which was believed to reflect intraoperative damage to the olfactory nerves. His visual acuity improved to 30/20 on the right and 20/200 on the left. Cerebrospinal fluid rhinorrhea resolved postoperatively. Magnetic resonance images obtained 48 months after surgery demonstrated no residual or recurrent tumor (lower right).

**Pathological Findings.** The tumor consisted of benign fibrous tissue with a varying mineralized content (Fig. 2). Some portions contained lamellar bone rimmed by osteoblasts, and other portions contained irregular trabeculae of woven bone without regular cement lines and osteoblastic cell lining. The histological diagnosis was a benign fibroosseous lesion.

**Case 2**

**History.** This 11-year-old boy was admitted with a 1-year history of nasal obstruction and headache.

Examination. On admission, the patient had no neurological deficits. Computerized tomography and MR images revealed a large multilobulated tumor involving the nasal cavity, sphenoid bone, and skull base (Fig. 3). The tumor had an irregular shape with a surrounding shell and thin internal septa. The shell and septa enhanced after administration of gadolinium. A biopsy specimen was obtained via a transnasal approach, and the cyst content was evacuated. The wall consisted of thin bone and fibrous membrane, and the cyst contained blood. The clinical and histopathological diagnosis was aneurysmal bone cyst.

Operation. The operative approach was the same as used in Case 1. Through a bifrontal craniotomy, frontonasoorbital and cribriform osteotomies were made. Both optic canals were released by means of an epidural approach. The tumor had thick fibrous walls covered with a thin layer of bone. Some of the bony walls were brown and granular, like an ossifying fibroma. Cyst contents consisted of blood. The tumor was totally removed. The skull base was reconstructed using a bipedicled temporoparietal galeal flap.

**Postoperative Course.** The postoperative course was uneventful. Olfactory function was well preserved. Magnetic resonance images obtained 40 months after surgery.
demonstrated total removal of the tumor without recurrence (Fig. 3 right).

Pathological Findings. The tumor showed various histological features of a benign fibroosseous lesion (Fig. 4). The fibrous wall of the blood-filled space was bordered by endothelial-like cells and multinucleated giant cells in connective tissue, a feature of aneurysmal bone cyst. Bony shells consisted of benign fibrous tissue and lamellar bone rimmed by osteoblasts, and features of ossifying fibroma were also seen. In the periphery of the lesion, such as the anterior clinoid process, the tumor showed a pattern of psammomatoid ossifying fibroma containing numerous small psammomatoid ossicles within a relatively cellular fibrous stroma.10,13

Discussion

The entity known as a benign fibroosseous lesion encompasses fibrous dysplasia and ossifying fibroma.22 These two lesions have been viewed separately: fibrous dysplasia, as an arrest in bone maturation in the non-lamellar or woven stage, whereas ossifying fibroma represents a true benign neoplasm of bone with lamellar bone formation and osteoblastic rimming.16 However, histological differentiation between them is difficult: many overlaps have been observed between specimens and within individual specimens.20 These varied lesions represent points on a spectrum of benign fibroosseous lesions.11 Aneurysmal bone cyst is a benign lesion of obscure pathogenesis, usually found in patients younger than 20 years old.48 The lesions grow rapidly, with a symptom duration averaging 3 months.4 The affected bone expands and “balloons” with many communicating cavities containing venous blood.1 Microscopic examination discloses several blood-filled channels bordered by a thin layer of spindle-shaped endothelial-like cells, surrounded by connective tissue containing numerous multinucleated giant cells.12 Hemosiderin-laden macrophages and new bone formation are also found within the stromal matrix.4 Aneurysmal bone cyst most likely represents a degenerative process associated with other primary bone lesions.6,13 Eversole2 reported 64 ossifying fibromas in mandibles or maxillae and found an aneurysmal bone cyst component in three cases. In another report, 21% of aneurysmal bone cysts in the jaws had developed in association with various primary bone lesions including fibroosseous lesions.5

In the literature, 12 reports of cases of benign fibroosseous lesions in the midline skull base include descriptions of symptoms.5,10,14,17,19,22 The most frequent symptoms result from mass effect (75%), such as exophthalmos (50%), nasal obstruction (33%), or local mass (25%). Other symptoms include disturbances of ocular movements (33%), visual loss (25%), and anosmia (8%). Meningitis or pneumocephalus such as in our Case 1 is a rare manifestation, because these lesions usually do not penetrate the dura mater. We have found only one reported case of ossifying fibroma with cerebrospinal fluid rhinorrhea and meningitis,17 in which a lesion in the anterior cranial fossa was histologically benign but repeatedly recurred with extensive invasion after partial removal. Ossifying fibroma is a steadily growing tumor that is more aggressive than fibrous dysplasia and recurs frequently in 21 to 44% of cases.27,20 After subtotal removal, regrowth is rapid, requiring more extensive surgery.3,10 Ossifying fibromas of the midfacial region and paranasal sinuses are more aggressive than those in the mandible and require wide local excision.16,17 The reported recurrence rate for aneurysmal bone cyst is also high, at 26%.6 For these lesions, wide exposure and complete excision represent the treatment of choice.2,4

Midline skull base lesions have been removed via a transcranial–transbasal approach16 or transfacial approaches3 such as transsphenoidal, Le Fort I, transoral, or transpalatal. The transcranial approach is better for excision of large lesions because the transfacial approach offers only a narrow corridor restricted by the pterygoid plates. The major drawback of the transcranial approach has been damage to olfactory function. The anterior craniofacial approach described by Spetzler, et al.,21 has solved the problem, providing a wide operative field while preserving olfactory function. This approach allowed us to reach an extensive area of the midline skull base from the planum sphenoidale to the clivus and the medial portion of the middle cranial base. This approach minimized trauma to important structures such as the optic nerves, sella, and cavernous sinuses, which were identified early in the procedure. Using this approach, we were able to achieve total excision of huge lesions in our cases. The anterior craniofacial approach is an effective method for treating benign fibroosseous lesions in the midline skull base, which should be resected totally.

References

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Fig. 4. Case 2. Photomicrographs of the surgical specimen showing features of aneurysmal bone cyst (upper), ossifying fibroma (center), and psammomatoid ossifying fibroma (lower). *Upper:* Fibrous wall of a blood-filled space bordered by endothelial-like cells and multinucleated giant cells. Original magnification × 200. *Center:* Bony shells consisting of benign fibrous tissue and lamellar bone rimmed by osteoblasts. Original magnification × 100. *Lower:* Numerous small psammomatoid ossicles embedded in cellular fibrous stroma. H & E, original magnification × 200.

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Address reprint requests to: Kiyoshi Saito, M.D., Department of Neurosurgery, Nagoya University School of Medicine, 65 Tsurumai, Showa-ku, Nagoya 466, Japan.