Neurosurgery: Official Journal of the Congress of Neurological Surgeons

March 1994, Volume 34, Number 3

Page 544

Case Report

Fronto-orbital Sphenoidal Fibrous Dysplasia

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WE REPORT THE clinical, radiological, and histological features of an 8-year-old boy with an unusual presentation of fronto-orbital sphenoidal fibrous dysplasia. The various forms of fibrous dysplasia are outlined and the differential diagnosis discussed. An approach to surgical management is proposed.
(Neurosurgery 34: 544-547, 1994)

Quick Access Outline

Key words: Cranial base tumor; Fibrous dysplasia; Spheno-orbital disease

A case of fronto-orbital sphenoidal fibrous dysplasia in an 8-year-old boy is described. The importance of this case lies in its unusual radiological and histopathological features. Operative management is detailed, and the classical types of fibrous dysplasia are discussed.

Craniofacial dysplasia is a benign disease of unknown origin, although some consider that a congenital

anomaly of mesenchymal development may be responsible. Craniofacial dysplasia represents approximately 2.5% of all bone tumors and 7.5% of benign bone tumors (5). It usually presents in childhood or adolescence and tends to stabilize after puberty. A recurrence in adulthood has been noted in about 37% of cases (3). The presence of fibrous connective tissue that contains poorly formed bone trabeculae without an internal lamellar structure is characteristic, the coarse woven bone being formed by direct metaplasia of the connective tissue.

CASE REPORT outlin

H.Q., an 8-year-old boy, was admitted in April 1992 with impaired vision in the left eye that was incidentally discovered during a consultation for rhinitis. In his history, repeated episodes of epistaxis were noted. His family history was unremarkable. An ophthalmological examination revealed reduced left visual acuity to hand movements at 20 cm and left optic atrophy. His right visual acuity was within normal limits. There were no other neurological deficits. There were no signs of precocious puberty.

Computed tomography (CT} revealed a well-defined hyperdensity confined to the left optic canal, extending toward the orbital roof and cerebral tissue and invading the sphenoidal sinus. Central and peripheral eggshell-like calcifications were found as well as thickening of the right anterior clinoid process (*Fig.1*).

Magnetic resonance imaging showed a nodular lesion, isointense on T1-weighted images, with strong uptake of gadolinium diethylene triamine-pentaacetic acid. It confirmed intracanalicular optic nerve compression and extension toward the orbital roof and into the sphenoidal sinus (*Fig. 2*).

Surgery was performed on April 17, 1992, through a combined extradural and intradural subfrontal approach. A grayish, firm, markedly hemorrhagic, fleshy tumor, very adherent to the dura mater, was found. The lesion was divided into smaller pieces, which then underwent curettage. The adherent dura mater was also excised. To release the optic nerve, dysplastic bone around the optic canal and left anterior clinoid process was removed by a high-speed airtome with a diamond tip until normal bone was reached. The intrasphenoidal part of the tumor was excised totally. A double plasty of the dura mater was performed to prevent leakage of cerebrospinal fluid.

The bony defect of the planum sphenoidale was reconstructed with a temporal bone graft. As the medial wall of the optic canal had been removed completely, the graft was inserted into the sphenoidal sinus, using the medial septum for support. The immediate postoperative course was uneventful.

The pathological examination confirmed the diagnosis of fibrous dysplasia (*Fig.3*). The microscopic examination of tissue fragments obtained during surgery showed intense proliferation of spindle-shaped fibroblasts with scant cytoplasm and long, fine, regular nuclei, which were organized in bundles embedded in an irregular collagenous stroma. In this fibrous connective tissue, multiple irregular trabeculae with occasional peripheral osteoblasts were observed. No lamellar structure was found, but poorly organized woven bone was. No inflammatory infiltrate or hemorrhagic features were seen. These findings are normally present in fibrous dysplasia.

At the time of discharge, the ophthalmological examination showed no improvement of visual acuity. Two weeks later, the patient was readmitted complaining of severe headaches and cervical pain. His neurological examination was within normal limits. A CT scan revealed the presence of a diffuse pneumocephalus indicating the presence of a dural defect.

Through a rhinoseptal approach, a bone graft was placed in a horizontal position and secured by a big piece of fatty tissue from the abdominal wall. The sphenoidal sinus was closed with two fragments of bone extracted from the nasal septum.

No cerebrospinal leakage leak occurred postoperatively. An ophthalmological examination 3 months later showed no improvement in his visual acuity. A second CT scan with bone windows showed the complete excision of the lesion, the disappearance of the pneumocephalus, and the good position of the bone graft. It also confirmed thickening of the right anterior clinoid process, probably secondary to dysplastic change (*Fig. 4*). Further clinical and radiological follow-up is needed.

DISCUSSION outlin

Clinical presentation outlin

Fibrous dysplasia of the skull represents 10 to 27% of monostotic cases (5,9) and occurs in all severe polyostotic forms (1,6). It causes considerable deformity, but malignant transformation appears in only 1% of cases (range, 0.4 to 4%) (2,4,5). Malignant change occurs more often after radiation in up to 44% of cases (5).

The most common presenting symptoms of craniofacial dysplasia are focal cranial swelling and exophthalmos or visual disturbances. Impaired vision is present in 20 (5) to 35% (11) of cases. Usually, it is progressive but can be acute after intratumoral bleeding (5) and is associated with cranial deformity in about 65% of cases (4,5,7). In our case, severely impaired visual acuity was an incidental finding and was not associated with any cranial deformity.

Radiological findings outlin

Radiologically, three forms of fibrous dysplasia of the skull are described (5). 1. *A compact form*. This is found in 50% of cases. The presence of homogeneous, increased density with thickening of bone gives a relatively radiopaque appearance. These lesions are found essentially at the base of the skull and involve the sphenoid body, the orbital roof, and the lesser wing of the sphenoid. CT shows a uniformly increased density. 2. *A lytic form*. This affects the cranial vault more frequently, with only 5% of cases showing extensive involvement of the cranial base. The anterior and medial base are transformed into a radiolucent area, and the sella is displaced upward and backward. Almost all the facial bones are involved. CT shows an irregular lucent area surrounded by a margin of increased density. No contrast enhancement is noted. 3. *A mixed form*. This is marked by the presence of areas of radiopacity alternating with radiolucent areas.

The radiological appearance of our case cannot be included in any of the three forms mentioned above. A CT scan demonstrated an area of increased density with striking contrast enhancement. Magnetic resonance imaging showed an isointense lesion on T1-weighted images and clear uptake of gadolinium diethylene triamine-pentaacetic acid.

Differential diagnosis outlin

The soft tissue appearance of a tumor associated with hyperostosis raises other possible differential diagnoses. 1. *Chordoma*. This rare tumor arises from notochordal remnants in the clivus and causes destruction of the basisphenoid. The most common initial complaint is diplopia. Although its clinical course was different, the lytic appearance associated with calcifications and a soft tissue mass in our case made chordoma a possible diagnosis. 2. *Chondroma*. A tumor of cartilage that arises from the spheno-occipital synchondrosis. The radiological appearance is that of a radiolucent lesion of the base of the skull with well-defined margins, often with adjacent areas of stippled calcifications. In our case, calcifications were noted, but the localization of the tumor was uncommon for a chondroma. 3. *Hyperostosing "en plaque" meningioma*. This needed to be considered in view of the bone thickening present and the soft tissue mass, but meningioma is rare in childhood, more common in female patients, and is usually located laterally on the sphenoid ridge and seldom in the sphenoidal sinus. 4. *A tumor of mucous membranes*. The presence of an intrasphenoidal mass made the diagnosis of a mucosal tumor possible, although this is very rare in childhood. 5. *Fibrous dysplasia*. The diagnosis of fibrous dysplasia was considered, although in the literature, it is emphasized that fibrous dysplasia is not associated with a fleshy intradural tumor (5).

Surgical approach outlin

A combined subdural and epidural surgical approach was necessary to ensure complete tumor excision and total decompression of the optic nerve. As already pointed out by Derome and Visot (5), the removal of the inferior wall of the optic canal is very difficult. Nevertheless, the removal of this wall was essential for the complete release of the whole optic nerve. As the ophthalmic artery is intimately attached to the inferior surface of the optic nerve, great care is needed in dissecting this area. In our case, injury of the ophthalmic artery was initially suspected because of severe bleeding, which was in fact arising from the tumor itself. Despite a double plasty of the dura mater, complete closure was not achieved and another operation through a rhinoseptal approach was required.

Histological findings outlin

Histologically, two macroscopic types have been described: a compact form, characterized by the presence of homogenous increased density with thickening of bone and sparse connective tissue, and a cystic form, with one or more cavities limited by a peripheral sclerotic eggshell-like margin. These cavities are separated from each other by fibrous structures and contain a browny-yellowish liquid. Sometimes these two forms occur simultaneously.

The microscopic appearance of these forms is unique. Poorly oriented trabeculae of bone are separated by cellular fibrous connective tissue. The bone appears woven rather than lamellar. Valls and associates, quoted by Spjut et al. (10), have demonstrated the formation of trabeculae from what they term *reticulohistiocytic* elements of the tumor. These bony trabeculae are irregular in size, shape, and distribution. Osteoblasts can be found near the area of transition to normal bone.

Morphologically, it is difficult to distinguish between an ossifying fibroma and fibrous dysplasia. In ossifying fibroma, the lesion is formed by fibrous tissue that contains variable amounts of lamellar bone trabeculae that appear interconnected in a single section. In the periphery of these trabeculae, osteoblastic

elements can be seen (8).

In our case, in most of the fragments examined, immature, woven bone was present, which is characteristic of fibrous dysplasia. This histopathological diagnosis is further supported by the fact that the right anterior clinoid process showed radiological signs of fibrous dysplasia.

CONCLUSION outlin

Aside from the sclerotic and cystic forms of fibrous dysplasia already described in the literature, we found another fleshy tumoral form. Where the tumor invades the sphenoidal sinus, a subfrontal approach should be followed by a rhinoseptal approach to ensure complete closure and to avoid cerebrospinal fluid leakage.

ACKNOWLEDGMENT outlin

The authors wish to thank Dr. M.C. Grangeponte for her contribution to the pathological study.

Received, February 11, 1993.
Accepted, October 1, 1993.

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COMMENTS outlin

Jan et al. have described a unique case of fibrous dysplasia affecting the optic canal and the supporting structures around it. This is not an uncommon situation in the child and adolescent. The authors have pointed out the problems of differential diagnosis and described their surgical approach. Our technique in this situation is to excise the sphenoidal and ethmoidal mucosa and to pack the area with fascia and fat, subsequently reconstructing the floor of the midline anterior cranial fossa with bone and placing a duraplasty over this. This technique separates the extracranial from the intracranial cavity such that if infection does occur in the sphenoid sinus, a transsphenoidal approach serves the purpose of removal of the fat and fascia. In addition, it allows for the correction of a cerebrospinal leak. However, in our experience of pediatric cranial base surgery, this incidence is very low. The authors have described the differential diagnosis and the radiological appearance in a precise manner.

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Compromised visual loss is an unusual presentation for fibrous dysplasia. The most common presentation for fibrous dysplasia is usually an asymptomatic cosmetic deformity of the involved bone or an incidental radiological finding. Fibrous dysplasia that extends to the cranial base, however, has the potential for compromising not only orbital contents but also cranial nerves and even the optic chiasm

and the pituitary gland. The authors' case is unusual in demonstrating a hyperdense lesion that was homogeneously enhanced and that was confined to the left optic canal. At surgery, a very vascular, fleshy tumor adherent to the dura was encountered. Complete surgical management required a subfrontal and subsequent rhinoseptal approach to repair cerebrospinal fluid leakage.

Current cranial base surgical techniques allow a more radical approach to these difficult bony lesions and allow primary reconstruction of the defects (3). In the decompression of the optic nerve, some authors have advocated the use of rongeurs rather than high-speed drills to prevent the transmission of thermal and vibratory energy to the nerve (2). On the other hand, others have demonstrated not only the preservation but the actual restoration of vision in cases of significant osseous compression of the optic canals, as in osteopetrosis. In these cases, careful irrigation and cooling of the area being decompressed as well as the careful preservation of the ophthalmic vascular supply is absolutely critical (1).

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