Monostotic fibrous dysplasia of the clivus

Case report

MICHAEL L. LEVY, M.D., THOMAS C. CHEN, M.D., AND MARTIN H. WEISS, M.D.

Department of Neurological Surgery, University of Southern California Medical School, Los Angeles, California

A case report of monostotic fibrous dysplasia of the clivus in a postadolescent woman is described. Although fibrous dysplasia of craniofacial structures is well documented, involvement of the clivus has not been reported. Diagnosis by clinical, radiographic, and histopathological features is detailed. Implications for the role of surgery and management are discussed.

KEY WORDS • fibrous dysplasia • transnasal biopsy • clivus

Craniofacial fibrous dysplasia is a benign disease representing approximately 3% of all bone tumors. It is one of multiple fibro-osseous lesions in which the normal matrix of bone is replaced with abnormal tissue, which may either contain osteoid or be calcified with collagen and fibroblasts. The etiology of fibrous dysplasia and its histological origins remain unclear. It most often presents in late childhood or early adolescence, and rarely appears in adults. The diagnosis of fibrous dysplasia is made from a combination of clinical, radiographic, and histopathological features.

We describe the first reported case of a monostotic fibrous dysplasia of the clivus. Diagnosis was obtained through radiographic evaluation and transnasal resection. The importance of conventional radiographic techniques in addition to radionuclide scintigraphic methods and magnetic resonance (MR) imaging is emphasized.

Case Report

This 37-year-old woman with a history of hypothyroidism presented with a 4-month history of severe intermittent headache.

Examination. On admission, the patient's physical and neurological examination were within normal limits. Clinical examination failed to reveal evidence of facial deformity. There was no tenderness on facial or paranasal palpation. Radiographic evaluation of the patient's skull revealed a dense, thickened clivus with cystic changes and aberrant areas of sclerosis. Computed tomography (CT) confirmed the presence of areas of radiolucency and sclerosis confined solely to the clivus, without evidence of extra-osseous extension. The jugular tubercle, temporal bone, sphenoid sinus, and cisterna interpeduncularis were free of involvement (Fig. 1). Radionuclide evaluation with 99mTc methyl diphosphonate-labeled red blood cells in a standard four-view scintigram confirmed involvement confined to the clivus without evidence of disseminated disease (Fig. 2). Magnetic resonance imaging revealed multiple high- and low-density regions on T1-weighted images confined to the clivus (Fig. 3).

Operation. The patient underwent a sublabial, transnasal approach for resection. A standard transnasal approach was taken with slight flexion of the head to allow for direct visualization of the clivus20 (Fig. 4). Access to the clivus was obtained by directing the dissection inferiorly along the vomer and the roof of the hard palate, with dissection of the mucosa of the nasopharynx from the clivus to gain operative access. Postoperatively, the patient experienced resolution of her headaches and has remained headache-free for 1 year.

Pathological Examination. The tissue obtained during surgery was fixed with 10% formalin and embedded within paraffin; 5-μm sections were made. Figure 5 presents a section prepared with hematoxylin and eosin staining; however, Ziehl-Neelsen, periodic acid-Schiff, Masson trichrome, and methenamine-silver stains were also performed. Pathological examination...
Monostotic fibrous dysplasia of the clivus

revealed fibrous connective tissue with islands of spindle cells. The normal marrow was replaced with foci of cartilage and metaplastic bone. Poorly formed and randomly arranged trabeculae representing coarse woven bone without internal lamellar structure were present throughout. Areas of hemorrhage, inflammatory reaction, and giant cells were noted.

Discussion

Fibrous dysplasia is a disturbance of postnatal development in which bone is replaced by fibrous tissue rather than resorbed. Typically, onset is early in childhood with progression into adolescence. Growth arrest occurs when skeletal growth ceases. It represents 2.5% of all bone tumors and 7% of benign tumors. There is a high incidence of involvement of the facial bones in blacks in addition to a female predilection. The age

Fig. 1. Computerized tomography scan, axial view, through the skull base. The areas of radiolucency and sclerosis are confined solely to the clivus.

Fig. 2. Technetium-99m methyl diphosphonate radionuclide evaluation confirming that involvement is confined to the clivus.

Fig. 3. Magnetic resonance image (TR 600 msec, TE 20 msec), sagittal view, demonstrating decreased signal in the central aspect of the clivus representing dysplastic fibrous tissue. Increased signal (arrow) represents replacement of normal cortex.

Fig. 4. Sublabial transnasal approach with slight flexion to allow for a direct approach to the clivus. (Modified with permission from Weiss MH: Transnasal transtemporal approach, in Apuzzo MLJ (ed): Surgery of the Third Ventricle. Baltimore: Williams & Wilkins, 1987, pp 476-494.)
at presentation ranges from 1 to 80 years. Initial presenting signs and symptoms occur in the first two decades in 83% of the patients with monostotic disease, and often consist of facial bone enlargement (80%). In patients with extracranial involvement, pain secondary to a pathological fracture is the most common complaint.

Disease Patterns

Predominant patterns include involvement of one bone (monostotic) or multiple bones (polyostotic). One example of polyostotic involvement is Albright's syndrome, which represents a constellation of polyostotic dysplasia, cutaneous hyperpigmentation, precocious puberty, and hyperthyroidism. Additionally, disseminated forms have been reported. Monostotic dysplasia is five times more common than polyostotic, and 30 times more common than Albright's syndrome. Craniofacial involvement has been described in the orbit, maxilla, zygoma, ethmoid, frontal, sphenoid, and temporal bones. Normal bone is replaced by collagen, fibroblasts, osteoid, and/or calcified tissue with subsequent symptoms reflecting specific bone involvement. Foraminal impingement can present as cranial neuropathies involving the first through fourth, sixth, and eighth cranial nerves. Histological origins are still unclear, with current theories ranging from microhemorrhage to failure of bone formation in embryonic mesenchyme.

Roenigenographic Studies

Fries described the roentgenographic features of the skull and facial bones in 39 patients with fibrous dysplasia. Pagetoid Type 1 characteristics are the most common (56%); this is notable for bone expansion and areas of radiopacity alternating with radiolucency (ground glass appearance). Pagetoid Type 2 occurs less often (23%), and has a homogeneous sclerotic appearance. Areas of radiolucency with well-defined margins occur in 21%, and are classified as Type 3. Evaluation by CT is useful in documenting the extent of bone involvement and possibly differentiating hyperostosis secondary to a meningioma or chonic sinusitis with maxillary involvement. Bone windows with thin cuts through the skull base are essential in describing foraminal involvement and cranial nerve compromise. Extracranial involvement is also well visualized on CT scans.

Radionuclide Evaluation

Radionuclide scintigraphic evaluation is an important tool for elucidating the presence of fibrous dysplasia. Given the hypervascularity of the involved bone, blood-pool scintigrams using 99mTc-labeled red blood cells will show increased uptake secondary to increased vascularity, osteoblastic reaction of bone, or deposition into calcified regions of tumor. Regular four-view studies are recommended in addition to vertical views. Radionuclide studies become positive when over 50% of calcified bone is replaced by fibrous tissue. They are more specific than CT for delineating the intra- and extraosseous extent of the lesion and demonstrating polyostotic or disseminated disease.

Histological Characteristics

Histologically, in fibrous dysplasia mature bone is replaced with immature woven bone with randomly spaced birefringent lines evident under polarized light. Osteoblasts and osteoclasts are notably absent. Tangled fibers can additionally become evident with silver reticulin stains. Hemorrhagic regions with associated inflammatory and giant-cell reactions are often described. The marrow is eventually replaced with fibrous connective tissue, islets of spindle cells, cartilage, and metaplastic bone. Multinucleated giant cells and foamy cells have also been reported. The bony trabeculae are hyperplastic and sporadic without evidence of normal internal laminar structure. The proportion of fibrous to osseous components can vary significantly. The cortex is often thinned and filled with fibrous tissue, depending on the extent of ossification and vascularity.

Diagnosis

In establishing a diagnosis of fibrous dysplasia, a combination of radiographic, radionuclide, and surgical evaluation is utilized to determine distinguishing features of these diseases from fibrous dysplasia. The differential diagnosis includes giant-cell granuloma, ameloblastic fibroma, odontogenic cyst, ameloblastoma, ossifying fibroma, and osteoma. Despite their similarity histologically, it is imperative that radiographic and histological differentiation of the juvenile and adult forms of ossifying fibroma be established, given the aggressive nature of the former when involving the maxilla. Biopsy is an integral aspect in obtaining and substantiating a diagnosis.
Monostotic fibrous dysplasia of the clivus

Management

The diagnosis of fibrous dysplasia is not an indication for surgical management. Usually small solitary lesions will remain static and not become symptomatic. Surgical management by excision and curettage is indicated when marked or progressive bone deformity, cranial nerve compromise, or pain syndromes become manifest. Delay of surgical intervention is recommended until adolescence or growth arrest occurs, unless neurological compromise becomes evident. Complete resection with extensive reconstruction when indicated is the treatment of choice. Ramsey, et al., reported a recurrence rate of 24% with incomplete resection. Edgerton, et al., suggested serial debridement and contouring until the lesion becomes static if complete resection is impossible. Hormone treatment has been reported to be ineffective, and adjuvant irradiation is contraindicated given the potential for malignant transformation. Chen and Fairholm reported 11 malignant transformations among 13 patients who received adjuvant irradiation. In our case, the diagnosis of monostotic fibrous dysplasia was confirmed at transnasal resection. Further intervention was not indicated.

Malignant Degeneration

The incidence of malignant transformation is unclear. Barat, et al., found 30 cases of malignant transformation in the literature up to 1983: osteogenic sarcoma occurred in 19 patients, fibrosarcoma in eight, and chondrosarcoma in three. They suggested that monostotic craniofacial lesions are most likely to become malignant (0.05% risk), with a mean interval of 13.5 years between diagnosis and transformation. Osseous transformation was 400 times higher following irradiation.

References


Manuscript received November 13, 1990. Accepted in final form April 30, 1990. Address reprint requests to: Martin H. Weiss, M.D., Department of Neurological Surgery, University of Southern California, 1200 North State Street, Suite 5036, Los Angeles, California 90033.