Journal of Korean Medical Science
Vol 7, No. 1, 58-61 March 1992

Ossifying Fibroma of the Sella Turcica

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Ossifying fibroma of the sellar turcica is extremely rare. There are only sporadic case reports in the literature. One such case simulating pituitary adenoma is presented in an 18-year-old girl.

Key Words: *Ossifying fibroma, Sella turcica, Pituitary adenoma*

INTRODUCTION

Pituitary adenomas and craniopharyngiomas are the usual lesions of the sellar region, while other tumors like chordoma, invasive nasopharyngeal carcinoma, neurofibroma, melanoma, and epidermoid cyst are rarely encountered. These all usually manifest as a pituitary adenoma, and their preoperative differentiations are, at times, difficult. Ossifying fibroma is a gradually expansile, well-marginated fibro-osseous lesion most commonly found in jaw bones. Its cranial occurrence is extremely uncommon as there are only a few case reports in the literature (Lehrer, 1969; Darsie and Kenan, 1971; Brette et al., 1987; Jammet et al., 1987; Zappia et al., 1990). Jawahar et al. in 1986 described a case of fibroma in the sellar region in a 13-year-old boy manifesting as bilateral primary optic atrophy. We here describe possibly the first case of ossifying fibroma of the sella turcica from India.

CASE REPORT

An 18-year-old girl was admitted complaining of gradual progressive dimness of vision in both eyes of one-year duration. There was no headache, or endocrinal imbalance.

Clinical examination revealed significant visual failure and bilateral primary optic atrophy. She was also found to have bitemporal hemianopia. There was no sensory-motor deficit. The plantars showed flexor response. A clinical diagnosis of nonsecretory pituitary adenoma was made.

A plain roentgenogram skull showed a ballooning of the sella turcica (Fig. 1). Cranial computed tomography demonstrated an enhancing high attenuating well-defined sellar mass with a surrounding area of low attenuation (Fig. 2). Carotid angiogram excluded the possibility of an internal carotid artery aneurysm.

A near total excision of the tumor was performed through a sublabial transphenoidal route. The postoperative phase was uneventful. Histopathological examination revealed a lamellar-type bone surrounded by cellular fibroblastic tissue with brisk osteoblastic activity (Fig. 3). With this picture a diagnosis of ossifying fibroma was made.

DISCUSSION

Many terms like cemento-ossifying fibroma, fibro-osseous lesion of the bone, osteofibroma, and fibro-osteoma are used to designate this condition, but the term ossifying fibroma popularized by Montgomery is the most acceptable. The term "ossifying fibroma" and "fibro-osteoma" are interchangable depending on whether the fibrous or bony tissue component predominates in the lesion. Menzel in 1872 mentioned the first case of ossifying fibroma involving the jaw bone as osteofibroma. Waldron and Giansanti emphasized that they are almost exclusively present in females, and more than 90% occur in the mandible. Hamner et al. found its high incidence in the maxilla, with no affinity to either sex. The cranial bones are the unfavorable sites (Takayama, 1985; Kuratsu et al., 1985; Kobayashi et al., 1986). Excluding the juvenile variety, they are seen in the third and fourth decades.

Although it is more common to see solitary lesions, multiple-site involvements do occur. They are confused clinically with bone cyst, fibrous dysplasia, nonossifying fibroma, and fibromyxoma. They present as diminution of vision, visual field defect, and primary optic atrophy like any sellar tumor. They can arise from the

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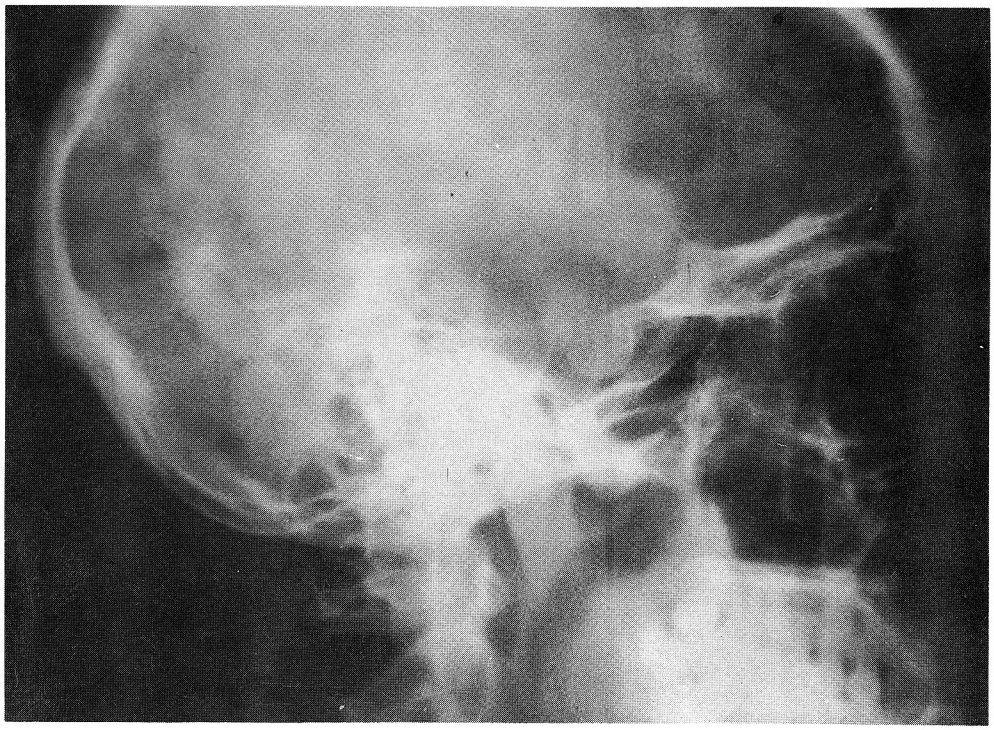

Fig. 1. Plain x-ray skull (lateral) shows widening and erosion of the sella turcica.

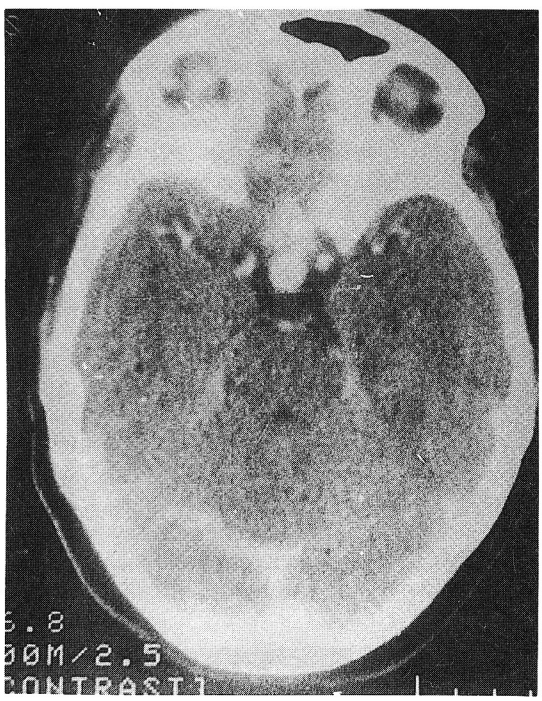

Fig. 2. CT scan head demonstrates the enhancing high attenuating mass in the sellar region.

pia arachnoid, dural fibroblasts, or from the perivascular connective tissue. It contains a uniformly cellular fibrous spindle cell growth arranged in a whorled or matted pattern with varying degrees of lamellar bone formation in different stages of maturation. It exhibits brisk osteoblastic activity. Capsule formation is an important feature to differentiate this pathology from fibrous dysplasia and reactive bone formation. Silver reticulin staining and polarizing light microscopy are useful adjuncts in histopathological examination, which demonstrate a parallel woven fiber pattern. In 1946, Billing and Ringertz distinguished 4 stages in the maturation process of ossifying fibroma.

Brette et al. (1987) documented the impossibility of histological differentiation between ossifying fibroma and monostic dysplasia with cranio-facial sites while Ye (1989) considered that fibrodysplasia and ossifying fibroma are different disease entities and should be distinguished by combination of clinical, radiologic and pathologic evidences.

The histological criteria for the diagnosis between the two diseases are: trabecular bone in a connective tissue with regular collagen fibers in the ossifying fibroma: nodular indented bony areas with irregular collagen frame in fibrous dysplasia. By histoenzymo-

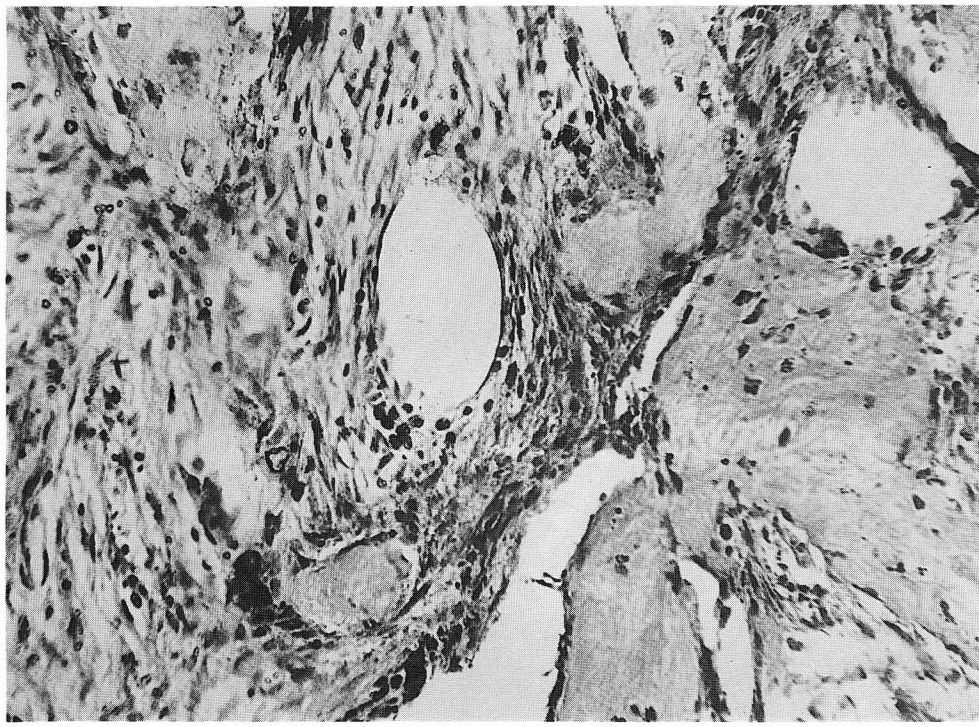

Fig. 3. Photomicrograph of the tumor reveals cellular fibroblastic tissue with brisk osteoblastic activity (H.E. × 140).

logy, the diagnosis between the two affections is not easy: same activity of ATPase and alkaline phosphatases in fibroblasts and osteoblasts. By electron microscopy, the morphology of the two lesions is different: in ossifying fibroma, numerous well-differentiated osteoblasts and large areas of ossification are seen: in fibrous dysplasia, undifferentiated cells are numerous and the collagen frame is irregularly mineralized (Chomette et al., 1987).

Radiologically, it appears as an osteolytic solitary lesion without periosteal reaction in the early stages, which later on becomes a well-circumscribed radiopaque lesion uniformly surrounded by a clearly-defined radiolucent rim. On CT; these lesions are expansile and circumscribed by a thick bony wall. Internal septations of bone density or enhancing soft-tissue density may be seen. CT scan is superior to conventional radiography in defining exact extent and site of lesions. Density of fibro-osseous conditions are variable due to the ratio of fibrous stroma and metaplastic bone present. Density measurements in fibrous dysplasia are 32-695 HU, in immature types of ossifying fibroma, consisting mainly of fibrous and osteoid tissue; 30-250 HU and could reach 690 HU, but definitively lower than normal bone (Irnberger, 1985). On MR, the bony walls are isointense with gray matter on T1-weighted images and are seen as areas of low intensity on T2-weighted images (Han et al., 1991). Differential diagnosis of benign fibro-osseous lesions include fibrous dysplasia, ossifying fibroma, osteoid osteoma, osteoblastoma, cementifying fibroma, florid osseous dysplasia, proliferative periostitis of Garre, focal sclerosing osteomyelitis and osteitis deformans (Antonelli, 1989), and they can be differentiated if clinical behavior, radiographic features, and hematologic changes are correlated with histologic picture.

Because the tumor tend to progress and can become clinically massive, early complete resection is advised whenever feasible to prevent clinically evident recurrences and potentially life-threatening complications (Zappia et al., 1990). Radiotherapy has been recommended following incomplete removal, but there is a potential risk of malignant transformation.

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