Case Report

Cementifying Fibroma of the Sphenoid Wing in a Child: A Case Report

N. K. Venkataramana, Shailesh A. V. Rao, N. Kirshna Chaitanya

Department of Neurosurgery, Brains Hospital, Bangalore, Karnataka, India

“Cementifying fibroma” is a benign tumor of the fibroblastic tissue containing masses of cementum-like calcified tissue, usually occurring between the third and fourth decades. Cementifying-ossifying fibromas are rare non-odontogenic, fibro-osseous tumors of the periodontal ligament that arise from the mesodermal germ layer. We report a 12-year-old male child, who presented with a swelling in the left temporal region near the angle of the left eye. These non-neoplastic, locally destructive tumors present as an osseous lesion involving the mandible, maxilla, zygoma, paranasal sinuses, orbit, and rarely the petromastoid regions. The preoperative diagnosis was not clear even with CT and MRI, and, hence, all fibro-osseous lesions were considered as differential diagnosis. The lesion was surgically resected completely, and the histology confirmed it as a cementifying fibroma. According to the WHO classification, this is a variant of cementifying fibromas, which represent a subgroup of cementomas, that is, fibro-osseous lesions containing cementum. Histologically, these are fibrous tissues with calcified structures resembling bone and cementum. Cranioplasty was done simultaneously, with successful clinical results.

**Keywords:** Cementifying fibroma, frontal bone, sphenoid wing

**Abstract**

Cementifying fibromas are rare, non-odontogenic equivalents of the ossifying fibroma of the periodontal ligament that arises from the mesodermal germ layer. This non-neoplastic, locally destructive tumor presents as an osseous lesion involving the mandible, maxilla, zygoma, paranasal sinuses, orbits, and, less frequently, the petromastoid region. The usual occurrence is in the third and fourth decade, with a preponderance in females. They are generally slow-growing benign tumors. Their occurrence in younger children is relatively rare and this subgroup can be classified as being of a juvenile variety. According to the World Health Organization classification, this tumor is a variant of cementifying fibromas, which represent a subgroup of cementomas, fibro-osseous lesions containing cementum. The benign fibro-osseous lesion can arise from any part of the facial skeleton and skull, often from membranous bones. Seventy percent of them occur in the head and neck region. Radical surgical resection is the treatment of choice. Only one case of cemento-ossifying fibroma of the petromastoid bone has been reported so far. We are reporting a case of cementifying fibroma involving the sphenoid bone presenting as localized growth of the skull. The diagnosis requires correlation of variety of clinical, radiological, and histological factors. The recurrence rate is high, particularly for lesions in the paranasal sinuses where the rate of recurrence is greater than 20%.

**Case Report**

A 12-year-old boy presented with a complaint of swelling over the left temporal region near the angle of the left eye and this had been noticed for more than one year. A trivial fall recognized the presence of such swelling, which was found to be increasing gradually.

**Address for correspondence:** Dr. N. K. Venkataramana, Brains Hospital, No. 560, 9th ‘A’ Main, Near Indiranagar Metro Station, Indiranagar, Bangalore 560038, Karnataka, India. E-mail: drnkvr@brains.org.in

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in size. There was no associated pain or any visual or neurological symptoms.

Clinical examination revealed a swelling involving the left temporal bone, extending into the lateral wall of the left orbit, pushing the eye ball slightly to the right. There was no similar swelling elsewhere in the body. Extra-axial proptosis of the left eye was observed with no restriction of eye movements. There were no pulsations or bruit. CT Brain revealed an intradiploic, hypodense expansile lesion involving the left sphenoid wing extending to the

**Figure 1:** Cementifying fibroma—radiological appearance, central lucency with peripheral sclerosis

**Figure 2:** Cementifying fibroma—compressing the left orbital structures causing fibrosis. No parenchymal involvement
lateral wall of the left orbit with an extracranial swelling. The lesion was measuring $4 \times 4.3 \text{cm}$ compressing the left orbital structures and causing mild proptosis. The central part of the lesion was radiolucent, with the surrounding sclerotic rim with well-defined margins. The outer table was expanded, causing a localized swelling [Figures 1–3]. All the four types of osteofibrous lesions in this location, such as ossifying fibroma, fibrous dysplasia, familial gigantiform cementoma, and cemento-osseous dysplasia, were considered as a differential diagnosis. However to relieve the compression on the orbital structures, we decided to resect the lesion.\textsuperscript{[13-25]}

**Surgical Management**

Left frontotemporal craniotomy was performed. The temporalis fascia and the muscle were intact. The swelling was found to be involving the bone and not infiltrating into any of the surrounding structures. The bone was cut with an oscillating drill, and the lesion was removed with a clear margin of the bone. At the end of the surgery, a significant part of the greater wing of sphenoid and the lateral wall of the orbit was removed, achieving gross total resection. The defect was reconstructed with cranioplasty by using a mayo-facial

![Figure 3: 3D Reconstruction showing a localized swelling](image)
Venkataramana, et al.: Cementifying fibroma of the sphenoid wing in a child

flap. The wound healed well, and postop CT showed no demonstrable lesion.[13-18]

**Histopathology**
Histopathology revealed a moderately cellular meningothelial cell tumor attached to the bone [Figure 4]. There were numerous calcified whorls [Figure 5]. Meningothelial cells did not show any atypical features. The tumor had a significant amount of cementum, justifying a diagnosis of cementifying fibroma.

**DISCUSSION**
Osseofibrous lesions of the skull are a rare group of heterogeneous disorders present as either a manifestation of a systemic skeletal disorder or a localized solitary process.[26] Cementifying fibromas are benign mesenchymal tumors belonging to this category. A cementifying fibroma is the odontogenic equivalent of the ossifying fibroma, which are clinically and histologically similar, if not identical.[1] It is a subtype of a broad category of congenital abnormalities of the bone formation known as craniotubular bone modeling disorders. There is a propensity to involve the skull base and once it reaches a sufficient size it compresses the neurovascular structures.[6,27] Neurological manifestations are often a result of compression of the cranial nerves in the exiting foramina at the skull base, and the most frequent nerve involved is the optic nerve.[28] Blindness from optic nerve compression is almost exclusively associated with the juvenile autosomal recessive form.[27] In contrast, in nomenclature by Kramer et al.[29] the cemento-ossifying fibroma is described as an osteogenic neoplasm and the fibrous dysplasia as a non-neoplastic bone lesion. The spectrum of possibility includes benign lesions as osteomas, osteoid osteomas, reactive expansile neoplastic lesions, primary malignant tumors, and metastatic lesions.[3]

In 1882, Menzel first described ossifying fibroma.[30] The exact etiology of this fibroma is not known. Trauma was suggested as a biological factor. However, developmental abnormality has also been proposed.[31] The recent World Health Organization classification of head and neck tumors in 2007 described four broad categories, which include osteofibroma, fibrous dysplasia, familial gigantiform cementoma, and cemento-osseous dysplasia.[32-36] Unlike fibrous dysplasia, the cemento-ossifying fibroma is well circumscribed from its surrounding bone and this lesion will continue to grow bigger, slowly and actively. Radiologically, 53% are radiolucent, 40% have a mixed radiolucent–radio opaque pattern, 7% have a mixed or mottled appearance,[37] and one case also had a radiolucent appearance. Another interesting feature is the centrifugal growth pattern; hence, these lesions grow, by expansion, equally in all directions. Thus, they currently have a localized swelling.[38] The treatment is total surgical resection. Large lesions reaching a size more than 80 mm in their greatest diameter have been termed “giant ossifying fibroma.”[26,39] The closest condition to be differentiated is fibrous dysplasia. Cementifying fibroma is round in shape with a dome-shaped expansion, whereas fibrous dysplasias are often elongated with a fusiform swelling. Margins are well defined in cementifying fibroma, whereas they are indistinguishable from the surrounding bone in fibrous dysplasia. Fibromas occur from 7 to 58 years of age, whereas the mean age of the presentation of fibrous dysplasia is 20 years. However, definite confirmation is done only with histological appearance. Juvenile (aggressive) ossifying fibroma was considered and excluded, because the histological description of

**Figure 4:** Numerous calcified whorls

**Figure 5:** Meningothelial cells do not show atypical features
this entity was significantly different from that of the current case.[40,41] Unlike the cemento-ossifying fibroma, the juvenile ossifying fibromas are aggressive, consisting of a cell-rich fibrous tissue with a cellular osteoid, woven bone, small foci of giant cells, and sometimes abundant osteoclasts related to the woven bone.[29] The juvenile ossifying fibroma occurs mainly during the second to the fourth decades of life, and it is more common in women than men.[5,27] Progressive cementifying osteofibromas need to be treated, as they have a benign osteoclasts related to the woven bone. Early surgical decision will help gross total removal.[42] Large lesions require cosmetic reasons and protection to the surrounding structures.[34-36] The recurrence rate is high, particularly for lesions in the paranasal sinuses where the rate of recurrence is greater than 20%.

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Conflicts of interest
There are no conflicts of interest.

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