*IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*
*e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 11 Ver. 6 (November. 2018), PP 69-74*
*www.iosrjournals.org*

# Intracerebral Primary Benign Chondroblastoma: Report of a Rare Case

Purwa R. Patil[1], Bhushan M. Warpe[2]*, Asif A. Khan[3], Shantilal Sisodiya[4]
*Corresponding Author: Bhushan M. Warpe*

***Abstract:*** *Chondroblasoma (CB) is a rare benign cartilaginous bone tumor arising from the epiphysis of long bones. Even rarer is intracerebralchondroblastoma. We report a unique case of primary benign chondroblastoma in left frontal lobe of cerebrum in 23 year-old male. He presented with intermittent frontal headache and vomiting of two weeks duration. Radio-imaging suggested the possibilities of meningioma and oligodendroglioma.The tumor was completely excised through left frontal craniotomy. Intra-operative squash smears revealed a benign, non-glial tumor with osteoclastic giant cells. Histologic examination showed typical features of benign chondroblastoma. On Immunostainingchondroblasts exhibited strong reactivity for S-100 protein and Vimentin. Osteoclastic giant cells were strongly positive for CD68. Tumor cells were negative for GFAP and EMA. To the best of our knowledge, this is the second case report of primary intracerebralchondroblastoma in the English literature. Here we discuss the differential diagnosis and probable histogenesis of intracerebral primary chondroblastoma.*

***Keywords:*** *Primary, benign, frontal lobe, Chondroblastoma, intracerebral, chondroblasts*

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Date of Submission: 13-11-2018 Date of acceptance: 28-11-2018

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## I. Introduction

Chondroblastoma (CB) or Codman tumor a rare, benign, cartilage forming bone tumorcomprises 1% of all primary bone neoplasm which usually occur in the epiphysis of long bones with only 1% seen in the skull.[1]It has been documented in different extra osseous locations including brain as sporadic case reports.[2-4]

We report a case of primary benign CB in left frontal lobe of cerebrum in a 24 year old male. To the best of our knowledge this is the second case report in world literature of primary benign CB occurring in the cerebrum.

## II. Case Report

A 23 year old right handed male laborer was admitted in our hospital with the complaints of severe and intermittent frontal headache and vomiting from 15 days and sudden onset, gradually progressing loss of vision in both eyes without diplopia since one week. There was no significant past medical history.

On neurological examination, higher functions were normal. There was no motor or sensory deficit but ophthalmic examination revealed reduced visual acuity, lateral gaze restriction on left side and pupil sluggishly reacting to light. Fundoscopic examination revealed pallor of temporal aspect of left optic disc. Also observed was left sixth nerve paresis.

CT scan of brain with and without contrast enhancement showed a hyperdense round space occupying lesion in the left frontal lobe with sharp margins and peripheral calcification. It showed peripheral bright contrast enhancement. It was interpreted as meningioma. MRI brain revealed a 7.3x5.2x5.4 cm large lobulated hypointense mass with hyperintense calcified component on T1 weighted images [Figure1]. The lesion showed mixed intensity on T2 weighted images. On post contrast study, thick peripheral enhancement was seen. There was no bony or dural attachment. Meningioma and oligodendroglioma were the differential diagnoses suggested on radiological findings.

The patient underwent left frontal craniotomy. On opening the dura, a lobulated calcified mass was seen reaching the surface near the frontal pole. Total excision of the tumor was done after dissecting it carefully from surrounding brain parenchyma. Intra-operatively possibility of brain tuberculoma was considered and the tissue was sent for squash cytologic examination.

95% ethyl alcohol fixed squash cytologic smears were stained with H&E stain. The smears were hypercellular and comprised of two types of cells in a non-fibrillary background. There were three dimensional sheets of polygonal cells having distinct cell borders, dense eosinophilic cytoplasm and round to oval, indented nuclei. Some of the nuclei showed nuclear grooves giving them coffee –bean appearance. The second component consisted of few osteoclastic giant cells with10 to 15, round nuclei [Figure 2]. There was no necrosis, psammoma bodies, intracytoplasmic hemosiderin pigment, inflammatory cells, intranuclear

pseudoinclusions and calcification. Thus possibility of tuberculoma or meningioma was less likely in this case and a diagnosis of benign non-glial tumor with osteoclastic giant cells was suggested.

Patient had a smooth post operative recovery. There were no focal sensory or motor neurologic deficits and total excision of tumor was confirmed on post operative CT scan.

In neuropathology division, a well circumscribed, lobulated, grayish white to red brown firm mass measuring 7x5x5 cm was received[Figure 3(a)]. Cut surface was grayish white to brown glistening with central cystic hemorrhagic area [Figure 3(b)].

H&E stained sections through the mass revealed a well circumscribed tumor comprising of lobules of polyhedral to spindle chondroblasts having distinct cell boundaries with dense eosinophilic to clear cytoplasm and round to oval, lobulated pale nuclei. Some nuclei had nuclear groove. Multinucleated osteoclastic giant cells were seen arranged singly or in small clusters in the stroma.

The mitotic activity was low. The most striking features were areas of chondroid differentiation, and chicken wire calcification between the degenerating cells. The calcification was also found in the cytoplasm of the cells especially in focal areas of heavy calcium deposition. Areas of secondary aneurysmal bone cyst formation were observed. A histologic diagnosis of CB was made [Figure 4 and Figure 5(a and b)]. The tumor showed diffuse S100 and vimentin positivity and was negative for EMA and GFAP. The osteoclastic giant cells were CD68 positive [Figure 6(a,b and c)].

On recent follow up, the patient is totally free of symptoms and doing well 11 months after the surgery.

## III. Discussion

Primary intracranial extraosseous cartilaginous tumors are extremely rare neoplasm and have been documented as isolated case reports in literature. They include both benign and malignant tumors such as chondroma, CB and chondrosarcoma.[2, 5-12]

Intracerebral CB is extremely rare tumor there being only one case report in the English literature in which authors have extensively discussed the radiological features and differential diagnosis of intracerebral [frontal lobe] CB.[2]

Intracerebral CB has to be distinguished from non-glial tumors showing focal chondroid differentiation, metaplastic cartilage formation, chicken-wire calcification, and ossification with haphazardly arranged osteoclastic type of giant cells in the stroma. The tumors showing these features are metaplastic meningioma (MM), CB like chondroma, chondroma, mesenchymal chondrosarcoma and clear cell chondrosarcoma (CCC).[5-16]

Intracerebral CB can be distinguished from MM with prominent ossification and calcification. Unlike MM, CB does not show dural tail on radiology. Microscopically MM is composed of cells with indistinct cell borders and oval nuclei with delicate nuclear chromatin and small nucleoli and pseudonuclear inclusions. The cells may be arranged in whorls, fascicular and syncytial pattern. A considerable element of fat, bone, cartilage or myxoid tissues is noted. It may show blood vessels of variable thickness ranging from small capillary like to dysplastic one having irregular thickness and focally myxoid features. These features are in contrast to the histologic features of CB. [13]

Histologically, the characteristic cells of CB are chondroblasts having uniformly, round to polygonal cells with well defined cytoplasmic borders, clear to dense eosinophilic cytoplasm and round to ovoid nuclei, arranged in pseudo-lobulated sheets. The nucleus usually displays clefts or longitudinal grooves and contains one or more small to inconspicuous nucleoli. Haphazardly distributed osteoclast-type giant cells are invariably present. Variably sized lobules of chondroid in the form of light-staining, amorphous, bluish to eosinophilic material accompany the chondroblasts.[16-18] Many cases show a fine network of pericellular "chicken wire calcification". Cytological atypia in the form of large, hyperchromatic nuclei may be observed in some chondroblasts, however such features do not adversely affect prognosis. Mitotic figures are observed but atypical forms are never noted. Areas resembling aneurysmal bone cyst may also be seen in 20 -25% cases of CBs. Ultrastructural studies reveal fetal chondroblasts showing deep nuclear grooves, abundant rough endoplasmic reticulum and long cytoplasmic processes while meningioma shows well formed desmosomes and interdigitating processes.[19]

Immunohistochemically chondroblasts are strongly positive for S100 protein, vimentin, SMA and cytokeratin. Multinucleate osteoclast- like giant cells shows CD-68 positivity. Reticulin stain reveal honeycomb pattern.

Although chondroblastoma like chondroma of soft tissue is not described in the cerebrum, it can be distinguished from intracerebral CB as the former shows features of CB such as hypercellularity, randomly distributed osteoclastic giant cells, lace like or chicken wire calcification and chondroid matrix which are focal in otherwise typical histologic setting of chondroma consisting of classical chondrocytes in a well defined lacunae and hyaline cartilaginous matrix.[14]

Chondroma is relatively less cellular lobulated tumor with abundant hyaline cartilaginous matrix as against mineralized fibrochondroid matrix with chicken wire calcification in CB. Also the chondrocytes with inconspicuous, small round nuclei and fine granular cytoplasm, always lie in lacunae.[16]

A mesenchymal chondrosarcoma of classical variety like CB is a well circumscribed tumor showing benign appearing chondroid areas. Foci of mature bone and calcification may be seen. However presence of diffuse sheets of highly undifferentiated small cells with perivascular clustering of cells rules out chondroblastoma.[10-12]

Clear Cell Chondrosarcoma (CCC)mimics CB, however the presence of clear chondrocytes with abundant cytoplasm with mild to moderate anaplasia and prominent osteoid and woven bone distinguishes CCC from CB. Immunohistochemistry does not help in distinguishing them as both show S100 and vimentin positivity.[15]

Total surgical resection of CB leads to cure, however it may recur locally or it may show metastasis especially to the lungs.[20-23] The histogenesis of intracerebral CB is unclear. As suggested in the literature for other chondroid lesions of CNS, the ectopic embryonic rests of cartilage cells, metaplasia of perivascular mesenchymal tissue, or traumatic displacement of cartilage elements can be considered as the probable cause of origin of CB.[5-7,24]

We report this case because of the extreme rarity of intracerebral CB. It should be one of the differential diagnoses during intra-operative frozen section /squash cytology of intracranial extraosseous cartilage forming tumors, as it carries a good prognosis and total excision of the tumor is often curative however regular follow up is essential as CBs in rare cases are known to undergo malignant transformation.[21,23]

**Figure with Legends:**

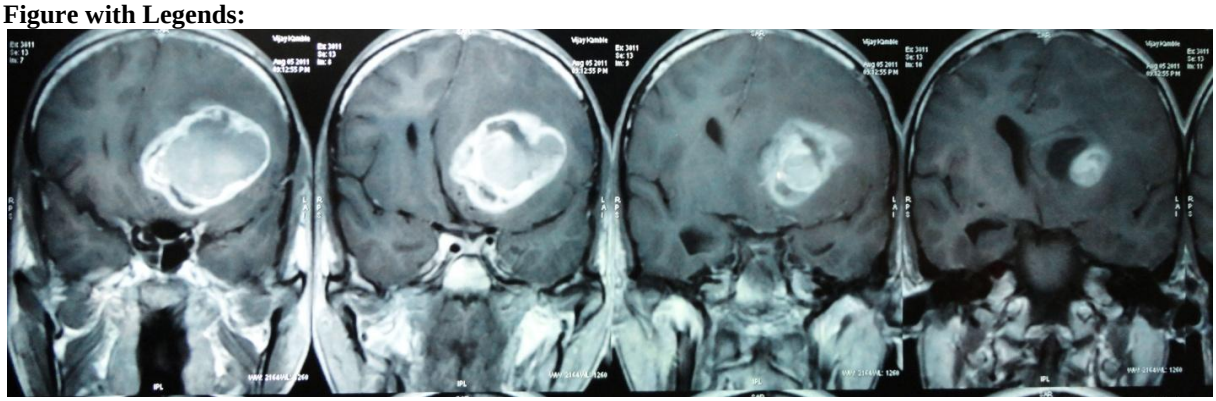

**Figure 1:** Coronal contrast enhanced MRI image showing a 7.3x5.2x5.4 cm large lobulated hypointense mass with hyperintense calcified component on T1 weighted images.

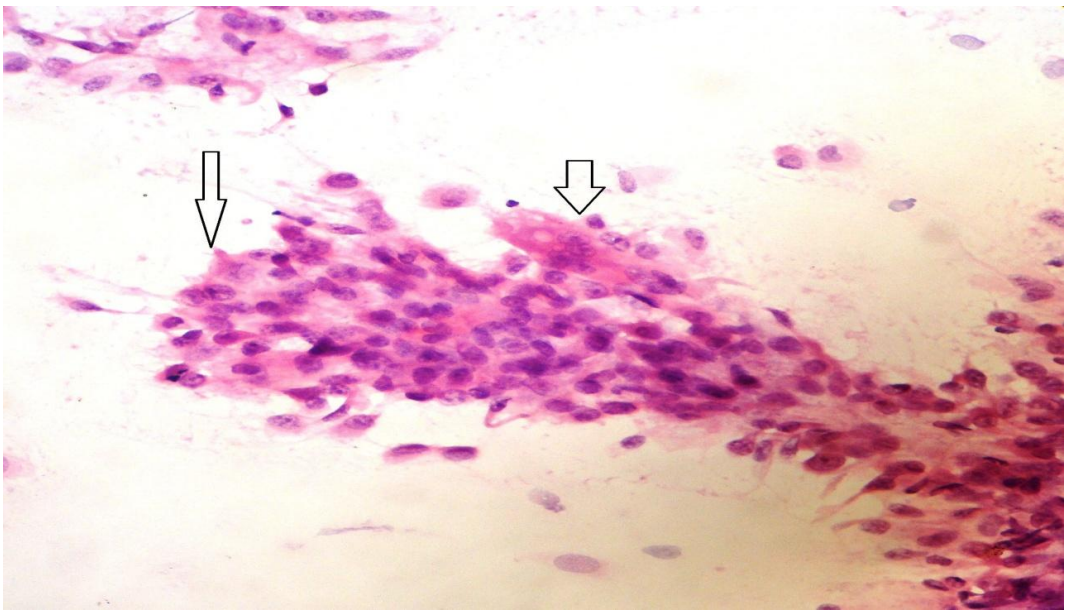

**Figure 2:** Photomicrograph of intra-operative squash cytology of chondroblastoma shows a hyper cellular smear with polygonal cells having distinct cytoplasmic borders, eosinophilic cytoplasm and round to oval, lobulated nuclei (large arrow); and osteoclastic giant cells (small arrow) (H & E, x200).

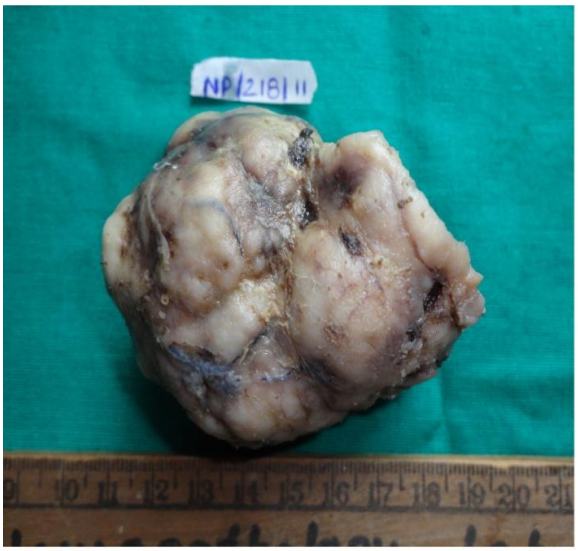

Fig. 3a

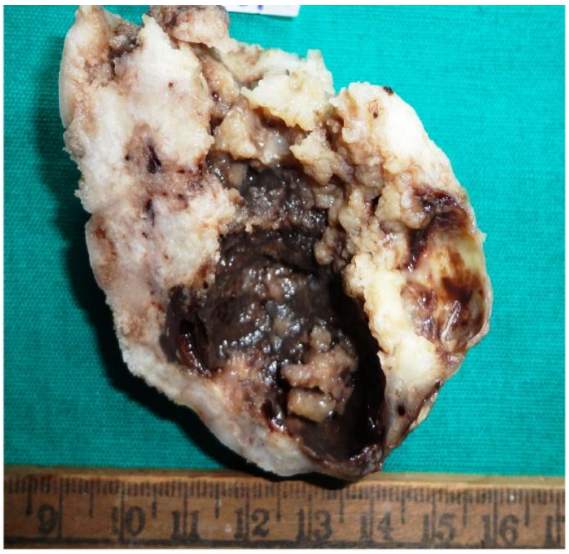

Fig. 3b

**Figure 3 (a,b):** Gross appearance of tumor reveals a well circumscribed white glistening mass measuring 7x5x5 cm with central cystic area.

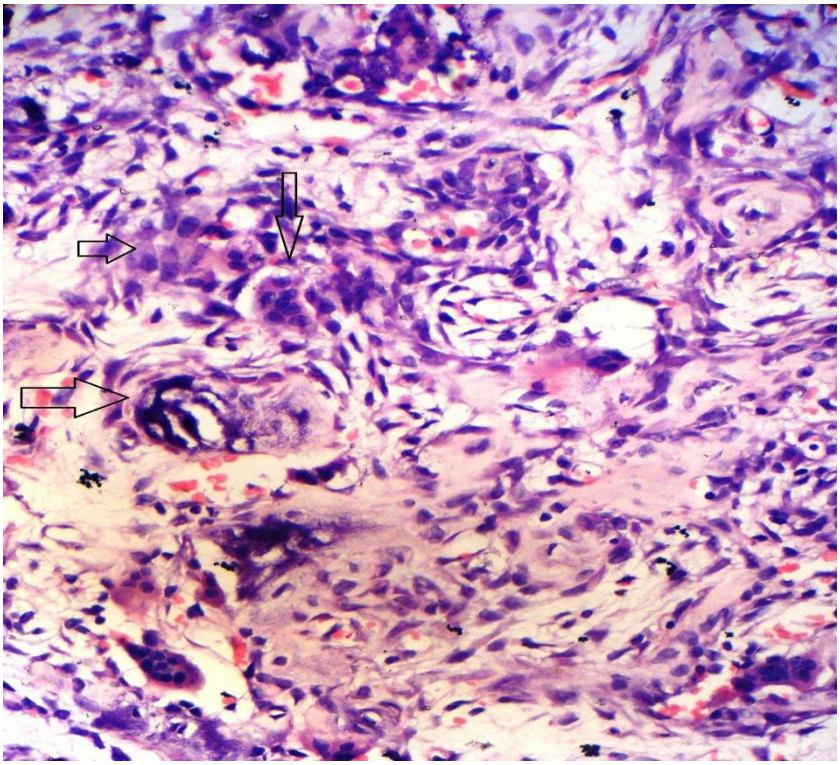

**Figure 4:** Photomicrographs shows mononuclear chondroblasts with nuclear grooves (small arrow), osteoclastic giant cells (medium sized arrow). Also reveal chicken-wire calcification seen (large arrow) (H & E, x400).

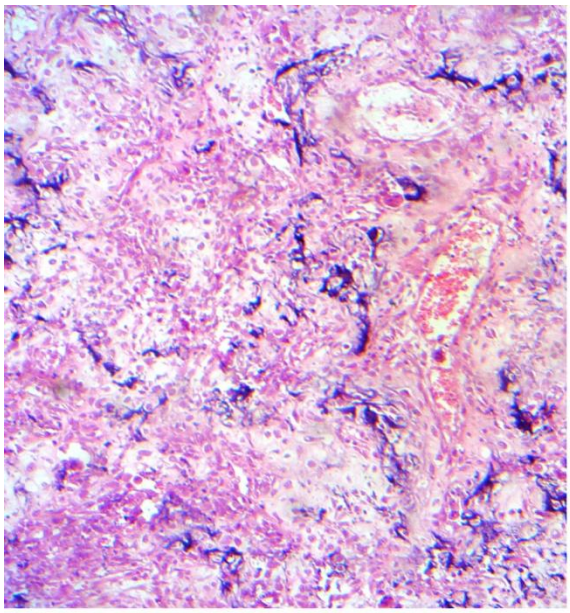

Fig 5a

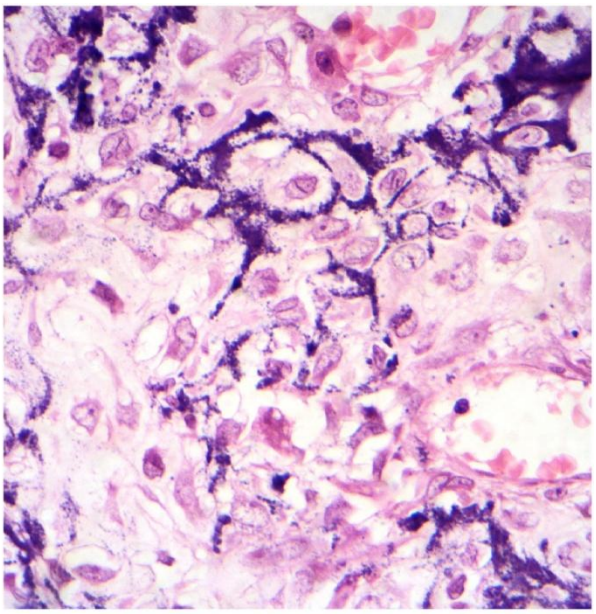

Fig 5b

**Figure 5:(a)** Photomicrograph shows areas of secondary aneurysmal bone cyst formation with tumor (H & E, x100). (b) Chicken-wire calcification amidst tumor cells that show nuclear grooving (H & E, x400).

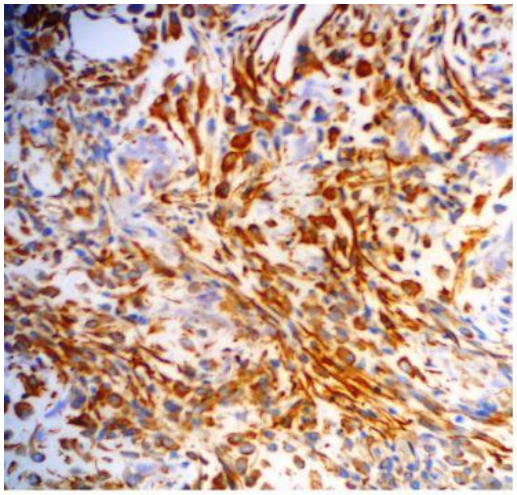

Fig 6a

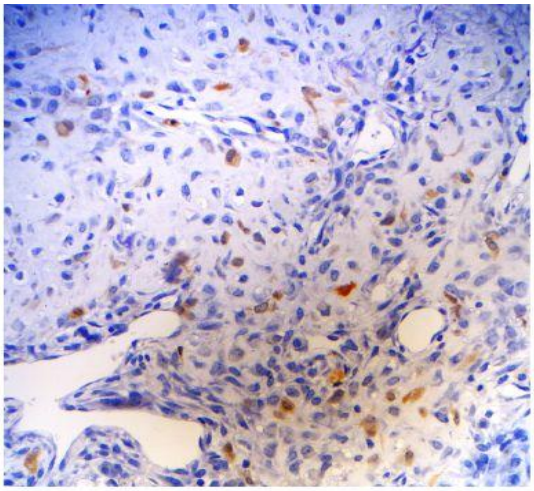

Fig 6b

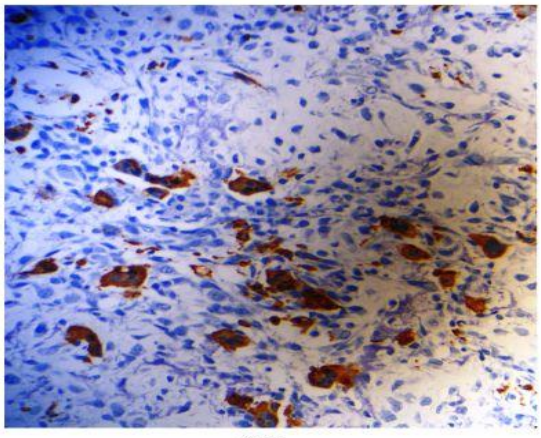

Fig 6c

**Figure 6:** Immunohistochemical staining showing (a) chondroblasts strongly positive for S100 protein (x200), (b) strong vimentin positivity of chondroblasts (x200), (c) CD68 positive osteoclastic giant cells (x200).

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Bhushan M. Warpe, "Intracerebral Primary Benign Chondroblastoma: Report of a Rare Case.". " IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17, no. 11, 2018, pp 69-74.