

Primary intracranial Parachordoma: An unusual tumor in brain

Rajesh K. Ghanta, Megha S. Uppin¹, Kalyan Koti², Monalisa Hui¹, Shantveer G. Uppin¹, Kanchan K. Mukherjee³

Departments of Neurosurgery, Suraksha Hospital, Vijayawada, ¹Pathology, Nizams institute of Medical sciences (NIMS), Hyderabad, ²Pathology, Dr Pinnamaneni Siddhartha Institute of Medical Sciences and Research Foundation (Dr PSIMS and RF), Chinoutpally, Krishna District, Andhra Pradesh, ³Neurosurgery, Post Graduate Institute of Medical Education and Research (PGIMER), Chandigarh, India

E-mail: *Rajesh K. Ghanta - ghantarajesh@yahoo.co.in; Megha S. Uppin - megha_harke@yahoo.co.in; Kalyan Koti - kalyan2305@yahoo.co.uk; Monalisa Hui - hui_monah@yahoo.co.in; Shantveer G. Uppin - drsguppin@yahoo.co.in; Kanchan K. Mukherjee - kk_mukherjee@hotmail.com

*Corresponding author

Received: 05 May 14 Accepted: 19 September 14 Published: 28 November 14

This article may be cited as:

Ghanta RK, Uppin MS, Koti K, Hui M, Uppin SG, Mukherjee KK. Primary intracranial Parachordoma: An unusual tumor in brain. *Surg Neurol Int* 2014;5:S506-11.

Available FREE in open access from: <http://www.surgicalneurologyint.com/text.asp?2014/5/15/506/145664>

Copyright: © 2014 Ghanta RK, This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Background: Parachordomas are rare soft tissue tumors commonly occurring in limbs, chest, Abdomen, and back. The World Health Organization (WHO) classification includes parachordomas in the same group as mixed tumors and myoepitheliomas. Exact histogenesis of this tumor is unclear.

Case Description: A 52-year-old male presented with headache and blurring of vision since one month. Preoperative computed tomography (CT) scan of brain revealed left parieto-occipital tumor extending up to the trigone. Total excision of the tumor was done. Histopathologically, the tumor was composed of relatively uniform cells with eosinophilic cytoplasm in a myxoid stroma and with cartilaginous and osseous metaplasia. The tumoral cells were immunoreactive for cytokeratin, epithelial membrane antigen (EMA), S-100, and vimentin. The constellation of findings revealed the tumor to be parachordoma. Magnetic resonance imaging (MRI) brain during follow-up at one year showed no recurrent tumor. No adjuvant therapy was given to this patient.

Conclusion: This is the first reported case of primary intracranial parachordoma. It is difficult to diagnose the lesion preoperatively by imaging alone. Long-term follow-up is necessary in view of few reports in literature of recurrence and metastasis, of parachordomas in other anatomical locations.

Key Words: Brain tumor, brain, intracranial parachordoma, parachordoma

Access this article online

Website:

www.surgicalneurologyint.com

DOI:

10.4103/2152-7806.145664

Quick Response Code:



INTRODUCTION

Parachordomas are rare soft tissue tumors. They commonly occur in extremities, the lower being more common than the upper.^[2] Parachordomas also occur in chest, back, and abdomen. They have been included in 2002 World Health Organization (WHO) classification of soft tissue tumors under the category of “tumors of uncertain differentiation” along with mixed tumors and myoepithelioma.^[7] They are

slow growing, benign lesions. A small number of these tumors develop local recurrences and are rarely seen to metastasize.^[16] We report the first case of an intracranial parachordoma in a 52-year-old male.

CASE REPORT

A 52-year-old male presented with history of headaches and blurring of vision since 1 month and right sided

weakness since 2 weeks. Clinical examination revealed normal higher mental functions, vision of 6/12 in both eyes without papilledema being present. Motor examination revealed mild right sided weakness-power 4/5 in both upper and lower limb. There was no prior medical history. The remainder of the physical examination was normal. Computed tomography (CT) scan of brain showed a tumor of size 3.5 × 3.2 × 2.9 cm

in the left parieto-occipital region extending up to the trigone [Figure 1]. The tumor was isodense and had punctuate calcifications. It appeared to be well defined with perilesional edema. Based on CT brain images, meningioma and choroid plexus tumor were considered as the differential diagnosis before surgery. Preoperative magnetic resonance imaging (MRI) brain with contrast imaging could not be done.



Figure 1: (a) Noncontrast CT scan of brain in axial plane reveal mass lesion in the left parieto- occipital region with dense calcification. (b) Noncontrast CT scan of brain in a more superior axial plane showing the tumor extending up to the trigone of left lateral ventricle with perilesional edema

A standard left parieto-occipital craniotomy was performed and the tumor was approached transcortically. Tumor was firm in consistency and total excision of the tumor was done. The tumor was gray-reddish in color, margins were well defined and a definitive plane between the tumor and surrounding brain was present. There was no infiltration into the adjacent brain parenchyma and the tumor was moderately vascular. There was no excess bleeding during the surgery. The histopathological examination of the tumor showed varied cellular morphology consisting of uniform oval to polygonal cells with moderate amount of eosinophilic cytoplasm arranged in nests, cords, and focally with adenoid configuration. The intervening stroma was fibrous and showed chondromyxoid areas with foci of osseous metaplasia [Figure 2a and b]. Immunohistochemistry: Tumor cells were positive for

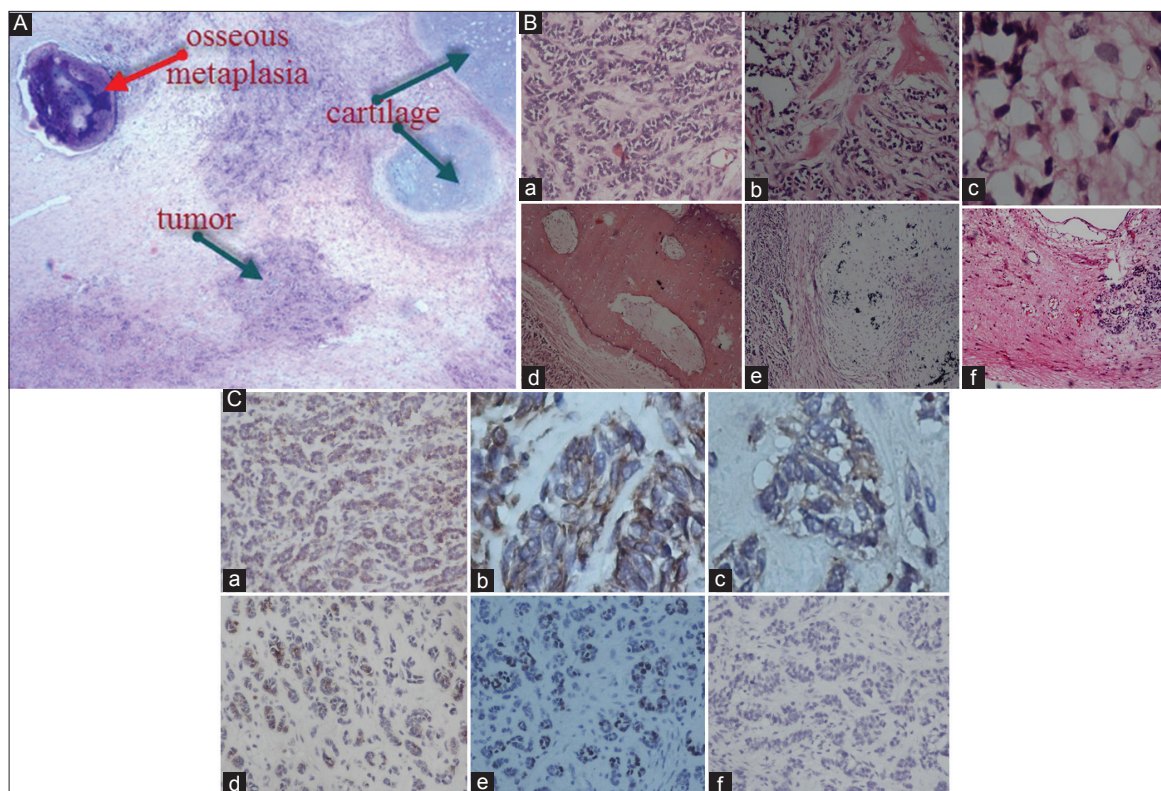


Figure 2: (A) Photomicrograph showing lobules of tumor with osseous and cartilaginous metaplasia. (Stain, hematoxylin and eosin; original magnification, ×200.) (B) (a-f) Photomicrograph showing tumor cells (a,b) arranged in the form of cords and trabeculae embedded in a hyalinized matrix, (c) focal physaliphorous like cells, (d,e) osseous and cartilaginous metaplasia, and (f) tumor infiltrating surrounding brain. (Stain, hematoxylin and eosin; original magnification, ×400.) (C) (a-f) Photomicrograph of the immunohistochemical panel showing tumor cell positivity for (a) pancytokeratin, (b) vimentin, (c) focal S-100 and (d) EMA. (e) Ki67 nuclear expression with a labeling index of 2% and (f) negative expression for GFAP. (Stain, diaminobenzidine ×400)

Pancytokeratin, epithelial membrane antigen (EMA), Vimentin, and S-100 and were negative for smooth muscle actin (SMA), Calponin, glial fibrillary acid protein (GFAP), Thyroid transcription factor (TTF), Cytokeratin 7 and 20, P63, and CD99 [Figure 2c]. The Ki 67 labeling index was 2%. The microscopic findings along with the immunohistochemistry favored a diagnosis of parachordoma. Karyotype analysis of chromosomes did not reveal any abnormality [Figure 3]. Whole exome genetic analysis was not done in this patient.

Postoperative course of the patient was uneventful. Since total excision of the tumor was done and the tumor having low proliferative index, no adjuvant therapy was given. Chest X-ray, ultrasound examination of the abdomen and screening MRI of the whole spine done 10 days after surgery did not reveal any abnormality. Follow-up contrast enhanced MRI scan of brain at one year follow-up, showed no recurrence of the tumor with adjacent gliotic changes [Figure 4]. A whole-body positron emission tomography (PET) CT scan at one year follow-up did not show any other lesion [Figure 5]. On follow-up at 16 months, his examination revealed 4+/5 power on right side and vision 6/12 in both eyes.

DISCUSSION

Parachordomas were first described by Lawskowski in 1951 as “chordoma periphericum.”^[3] The term parachordoma was first used by Dabska in his series of patients in 1977.^[3] Parachordoma is a rare soft tissue tumor of unknown origin occurring in the limbs, chest, back, and abdomen. Parachordomas have been reported to occur in the pelvis and gastric serosa also.^[16,17] Fewer than 60 cases of parachordoma have been reported till date,^[2,16] and include intraneural parachordoma^[4] and

parachordomas of the skull bone.^[19] However, there have been no reported cases of an intracranial parachordoma till the present reported case. Clabeaux *et al.* in a review of 45 cases of parachordomas in 2008 noted slight male predilection with the mean age of patients in their series being 34.4 years (range, 4–86 years).^[2]

The preoperative radiological diagnosis of intracranial parachordoma is difficult. In our case, based on the location and other CT characteristics of the lesion, we thought of choroid plexus papilloma and meningioma as differential diagnoses. Intraoperatively we found the tumor to be firm in consistency with no evidence of infiltration or involvement of the occipital horn of the left ventricle. Total excision of the lesion was possible as the tumor was well defined, encapsulated and after initial debulking of the tumor, surgical plane between the tumor and cerebral parenchyma facilitated the tumor to be removed completely. Postoperatively, chest X-ray, ultrasound abdomen, MRI of whole spine, PET scan of whole body were done to rule out a primary tumor. During follow-up, which these patients require for a long time, CT/MRI brain should be sufficient to rule out recurrent tumor.

Parachordoma was initially considered to be a chordoma in nonaxial location, but now it is considered as a unique entity with distinct immunohistochemical profile.^[2,5] Parachordoma is considered a slow growing tumor of an indolent, less aggressive nature than chordoma. The differential diagnosis for this tumor include extraskeletal myxoid chondrosarcoma, chordoid meningioma, and chordoma.^[11] Parachordomas histologically resemble mixed tumors of the salivary gland. There are three uniform-appearing cell types, namely epithelioid cells with eosinophilic to clear and vacuolated cytoplasm resembling physaliphorous cells, smaller glomoid cells, and spindle cells. Each cell type is present in varying proportions with arrangements in nests, cords, ductules, and are embedded in a hyalinized and chondromyxoid matrix. Parachordomas can show divergent differentiation in the form of squamous, adipocytic, osseous, and cartilaginous metaplasia. The tumor cells on immunohistochemistry show positive expression for S100 protein, Leu-7, keratin (CAM5.2), and EMA and negative staining for cytokeratin CK7 and CK19, carcinoembryonic antigen (CEA), muscle specific actin (MSA), SMA, desmin, GFAP, CD31, or CD34.^[6-8] The present case showed a similar histological appearance and staining pattern on immunohistochemistry as described earlier.

A definitive diagnosis is achieved using immunohistochemistry along with light microscopy. We have enumerated the immunohistochemistry findings for various tumors in Table 1. Chordoid meningioma can be composed of eosinophilic vacuolated cells in a myxoid matrix and can show prominent cartilaginous

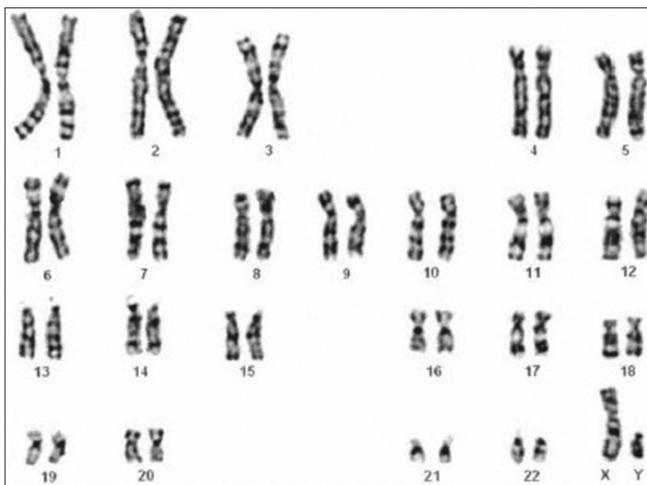


Figure 3: Chromosomal analysis of cultured peripheral blood lymphocytes. GTC – banded karyotypic analysis revealed no numerical or structural chromosome abnormalities and indicated a normal male karyotype

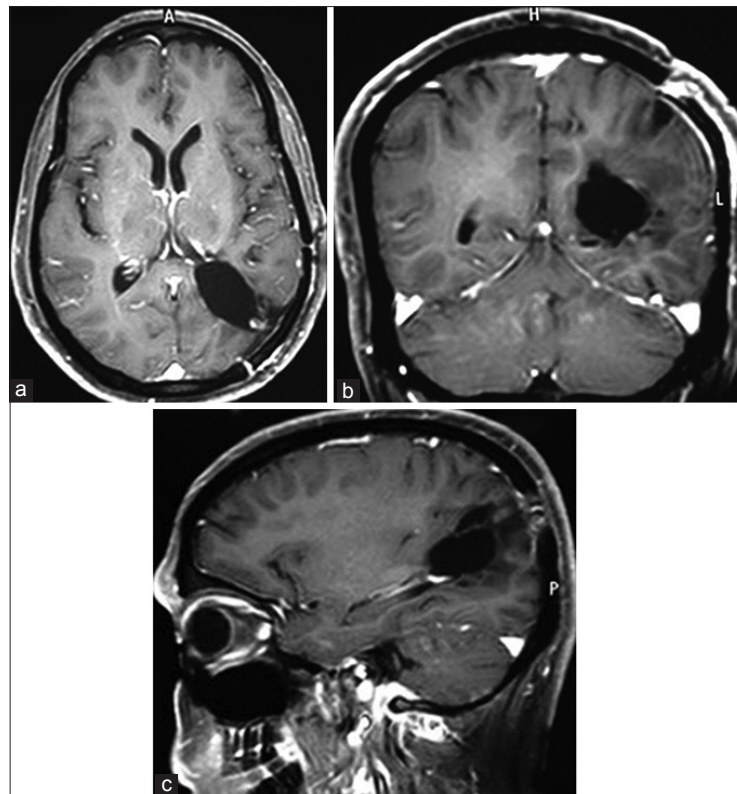


Figure 4: Contrast enhanced T1W MRI scan in axial plane (a), coronal plane (b), sagittal plane (c) at one year follow up showing completely resected tumor replaced by gliotic changes

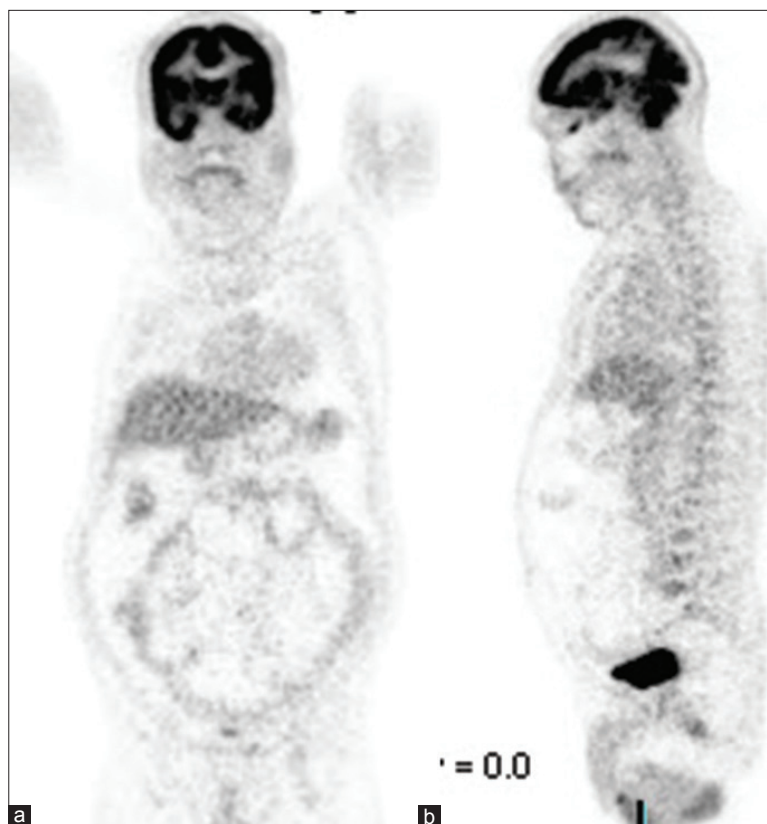


Figure 5: ^{18}F -fluorodeoxyglucose (FDG) PET/CT scan done at one year follow-up showing no evidence of intracranial or extracranial lesions. Fig 5a showing maximum intensity projection in coronal plane. (b) showing lateral view of FDG-PET scan

Table 1: Differential diagnosis of tumors resembling parachordoma based on immunohistochemistry

Tumor	S-100	Pancytokeratin	EMA	Vimentin	GFAP	Thyroid transcription factor	CK 7	Microtubule associated protein
Parachordoma	+	+	+	+				
Gliosarcoma				+	+			
Chondroid chordoma	+	+	+	+			+	
Chordoid meningioma			+	+				
Myxoid chondrosarcoma			+	+				+
Choroid plexus papilloma						+	+	

EMA: Epithelial membrane antigen, GFAP: Glial fibrillary acidic protein, CK 7: Cytokeratin 7

metaplasia.^[13] Otherwise a classic case of chordoid meningioma usually shows focal areas resembling the conventional transitional meningioma, which were not identified in our case. Choroid plexus papillomas can show unusual histologic features such as oncocyctic transformation, mucinous degeneration, xanthomatous change, glandular architecture, osseous, and cartilaginous metaplasia. However, the present tumor did not reveal any papillary areas, which is a classic histomorphology of choroid plexus papillomas.^[10] Chondroid chordoma is a variant of chordoma that also shows prominent areas of cartilage along with classical physaliphorous cells.^[6] The location of the tumor in the present case is not common for conventional or chondroid chordomas. The myxoid type of chondrosarcoma is composed of strings of rounded cells in a more or less myxoid matrix.^[15] There are only a few cases reporting the cytogenetics of parachordoma. Folpe *et al.* found trisomy 15 and loss of chromosome 1, 16, and 17 in one case with chromosomal studies.^[8] Tihy *et al.* reported a loss of chromosomes 9, 10, 20, and 22 in seven cells, a loss of chromosome 17 in four cells and a structural rearrangement of chromosome 3 [del (3q)] and chromosomes 2 and 4 [t (2p; 4q)] in a recurrent tumor.^[18]

The present treatment of parachordomas includes surgical resection with wide margins. The present existing literature on parachordomas does not advocate adjuvant therapy after resection with wide margins.^[2] Role of radiotherapy and chemotherapy is restricted to cases of parachordoma with metastasis.^[1] Parachordoma is a slow growing benign tumor with occasional late recurrences and rare instances of metastasis.^[2,4] Malignant parachordoma's have been reported in literature.^[11] Local recurrences and metastases are more frequent in patients with a histologically malignant appearance.^[11] Recurrence of the tumor was noted in 9 patients (20%) and metastasis in 5 patients (11.1%) in a review of 45 patients by Clabeaux *et al.*^[2] Recurrence as early as 3 months^[14] and as late as 12 years^[3] after initial treatment has been reported. Long-term follow-up of patients with

parachordoma is advisable in view of possibility of late recurrence. Metastasis of parachordoma has been reported in seven cases till now.^[2,4,9] Regional metastasis to distal metastasis can occur. Few cases of fatal parachordoma have been reported.^[1,12]

This is the first reported case of primary intracranial parachordoma. No adjuvant therapy was given in our patient as the natural history of this slow- growing tumor appears benign when a total excision of the tumor is performed and histopathology reveals a low proliferative index of tumor cells.

CONCLUSION

Parachordoma's are rare soft tissue tumors and we describe the first reported case of an intracranial parachordoma. There were no unusual factors associated with the resection and the authors advocate that patients diagnosed with this rare pathology require long-term and diligent follow-up in view of few reports in literature of recurrence and metastasis, of parachordomas in other anatomical locations.

ACKNOWLEDGMENTS

The authors would like to thank Dr. S. Kiran, for helping to provide general assistance in this work. The authors gratefully acknowledge HCG PET CT scan centre, Vijayawada and its director Dr. Gopichand M, Surgical Oncologist for their help in providing PET scan results and images for this work.

REFERENCES

1. Abe S, Imamura T, Harasawa A, Ishida T, Unno K, Tateishi A, *et al.* Parachordoma with multiple metastases. *J Comput Assist Tomogr* 2003;27:634-8.
2. Clabeaux J, Hojnowski L, Valente A, Damron TA. Case report: Parachordoma of soft tissues of the arm. *Clin Orthop Relat Res* 2008;466:1251-6.
3. Dabska M. Parachordoma: A new clinicopathologic entity. *Cancer* 1977;40:1586-92.
4. De Comas AM, Deavers MT, Raymond AK, Madewell JE, Lewis VO. Intraneural parachordoma of the arm with regional metastases. *Skeletal Radiol* 2011;40:943-6.
5. Fisher C. Parachordoma exists--but what is it? *Adv Anat Pathol* 2000;7:141-8.

6. Fisher C, Miettinen M. Parachordoma: A clinicopathologic and immunohistochemical study of four cases of an unusual soft tissue neoplasm. *Ann Diagn Pathol* 1997;1:3-10.
7. Fletcher CDM, Unni KK, Mertens F. *Pathology and Genetics of Tumours of Soft Tissue and Bone*. Lyon: IARC; 2002. p. 198-9.
8. Folpe AL, Agoff SN, Willis J, Weiss SW. Parachordoma is immunohistochemically and cytogenetically distinct from axial chordoma and extraskeletal myxoid chondrosarcoma. *Am J Surg Pathol* 1999;23:1059-67.
9. Guedes A, Barreto BG, Barreto LG, de Oliveira Araújo IB, Queiroz AC, Athanazio DA, et al. Metastatic parachordoma. *J Cutan Pathol* 2009;36:270-3.
10. Hasselblatt M, Jeibmann A, Guerry M, Senner V, Paulus W, McLendon RE. Choroid plexus papilloma with neuropil-like Islands. *Am J Surg Pathol* 2008;32:162-6.
11. Karakaya YA, Ozekinci S, Büyükbayram H, Mizrak B. Parachordoma: A recurrent case and review of the literature. *Türk Patoloji Derg* 2011;27:173-6.
12. Kinoshita G, Yasoshima H. Fatal parachordoma. *J Orthop Sci* 2007;12:101-6.
13. Matyja E, Grajkowska W, Łazarczyk M, Marchel A, Czernicki T. Chordoid meningiomas of a different histopathological pattern A report of two cases. *Folia Neuropathol* 2006;44:34-41.
14. Niezabitowski A, Limon J, Wasilewska A, Rys J, Lackowska B, Nedoszytko B. Parachordoma--a clinicopathologic, immunohistochemical, electron microscopic, flow cytometric, and cytogenetic study. *Gen Diagn Pathol* 1995;141:49-55.
15. Salaou O, Ibahioin K, Karkouri M, Kamar AE, Azhari AE. Intracerebral chondrosarcoma. *Pan Arab J Neurosurg* 2010;14:90-3.
16. Samaka RM, Kandil MA. Huge pelvic parachordoma: Fine needle aspiration cytology and histological differential diagnosis. *Rare Tumors* 2012;4:e53.
17. Spivach A, Zanconati F, Tirabosco R, Falconieri G. Parachordoma of the gastric serosa: Report of a myxoid mimicry in an unusual location. *Int J Surg Pathol* 2007;15:307-10.
18. Tihy F, Scott P, Russo P, Champagne M, Tabet JC, Lemieux N. Cytogenetic analysis of a parachordoma. *Cancer Genet Cytogenet* 1998;105:14-9.
19. Zhou F, Gong H, Jiang J, Zee CS, Wan H. Parachordoma of skull. *J Neuroradiol* 2010;37:247-8.