CASE REPORT

Plasmacytoma of the clivus

R. Amita, S. Sandhyamani, Suresh Nair[1], T. R. Kapilamoorthy[2]

Departments of Pathology, [1]Neurosurgery and [2]Imaging Sciences and Interventional Radiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India

ABSTRACT

We report a case of solitary plasmacytoma of the clivus in a 55 year old male.

Key words: Clivus, intracranial, plasmacytoma

Introduction

Plasmacytoma is a localized clonal proliferation of plasma cells arising in bone or soft tissue. It accounts for <1% of head and neck tumors. Plasmacytoma is classified by World Health Organization into solitary bone plasmacytoma (SBP) and extramedullary plasmacytoma (EMP).[1] The most common site is upper aerodigestive tract, 80% occurring in nasal cavity and nasopharynx. Solitary intracranial plasmacytoma is rare, with only close to 20 cases reported in the literature.[2] Most of the cases reported show involvement of the skull vault. Involvement of the base of the skull is rare with only a few exceptional reports citing clivus as the site.[3] We report a case of plasmacytoma of the clivus in a 55 years old male.

Case Report

A 55-year-old male presented with insidious onset diplopia while looking to the right and to distant objects of 3 months duration. There was no history of visual blurring, jaw deviation, facial asymmetry, hearing loss or tinnitus. There was no associated headache, vomiting, seizures or loss of consciousness, limb weakness, gait, and bowel or bladder disturbances. On examination, patient was conscious with stable vital signs. Higher mental functions were normal. There was right side sixth nerve palsy and nystagmus on looking to the right. Visual acuity and visual field examination were normal. There were no other cranial nerve, motor or sensory deficits. Other system examinations were within normal limits. Magnetic resonance imaging of the brain showed an expansile mass lesion involving entire clivus that was T1- and T2-isointense and showed contrast enhancement. There was erosion of posterior wall of the sphenoid sinus with lesion bulging into the sinus. Pituitary was normal [Figure 1]. With a preoperative diagnosis of clival chordoma infiltrating into the sphenoid sinus, the lesion was decompressed by subfrontal trans basal approach.

Microscopy showed respiratory epithelium with an underlying neoplasm composed of sheets of plasma cells with a moderate amount of cytoplasm and eccentric round nuclei [Figure 2]. Cells were positive for CD 138, CD 38 and CD 56, while negative for CD 20, cytokeratin and synaptophysin. Ki 67 (MIB) labeling index was <5%. Cells were kappa light chain positive and negative for lambda light chains thus establishing monoclonality. A diagnosis of plasmacytoma of clivus was given, and workup for myeloma was suggested.

Patient underwent whole-body X-ray studies, serum and urine electrophoresis, complete blood counts, serum calcium levels and a bone marrow biopsy, as part of myeloma investigations, which were negative.

Patient received radiotherapy (50 gray given in 25 fractions) and is now 8 months postoperative and doing well. The right side lateral rectus palsy is still persisting.

Discussion

Plasma cell tumors affecting the skull base generally appear as a manifestation of underlying multiple myeloma (MM). Solitary plasmacytoma of the skull base is rare with <10 reported cases of clival plasmacytoma in literature.[2-5] A brief summary of the cases is provided in Table 1. Clival plasmacytoma may represent an SBP or may be involved by EMP involving the submucosa of the sphenoid sinus as in our case.

Access this article online	
Quick Response Code:	Website: www.asianjns.org
	DOI: 10.4103/1793-5482.148788

Address for correspondence:
Dr. S. Sandhyamani, Department of Pathology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram - 695 011, Kerala, India.
E-mail: amita.rnair@sctimst.ac.in

By definition, patients with solitary plasmacytoma do not have evidence of underlying systemic disease at the time of diagnosis.[1]

In general, solitary intracranial plasmacytomas remain asymptomatic for a long period of time. The symptoms are related to compression of cranial nerves and headaches. Tumors in the clivus produce diplopia due to compression of the sixth cranial nerve, which has the longest path in this anatomic region. This was true in our case also.[2]

Other cranial nerves affected include II, V, VII and VIII, according to the order of frequency. Rare case reports of compression of the posterior inferior cerebellar artery leading to a lateral medullary and bulbar syndrome are also on record.[5]

A radiological differential of a lesion in clivus includes chordoma, chondrosarcoma, meningioma, invasive pituitary adenoma, lymphoma, metastasis, and osteosarcoma. Positron emission tomography with fluorodeoxyglucose has been found to aid in the detection of unsuspected sites of medullary and extramedullary disease.[5]

Due to the rarity of plasmacytoma at the skull base and the nonspecific clinical and radiological findings, in most cases

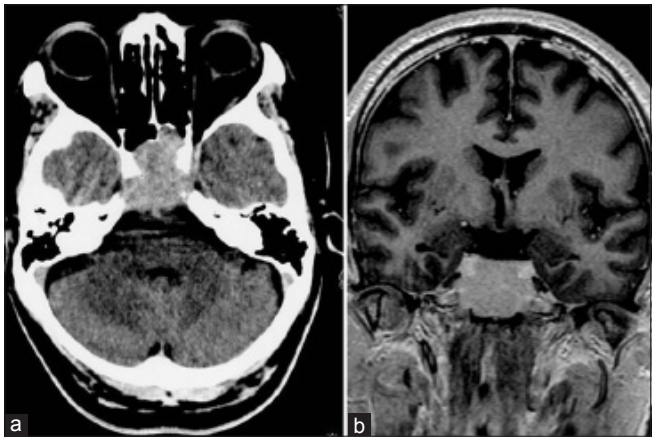

Figure 1: (a) Magnetic resonance imaging brain shows an expansile lesion involving entire clivus and eroding the posterior wall of sphenoid sinus. (b) Saggital view of the lesion

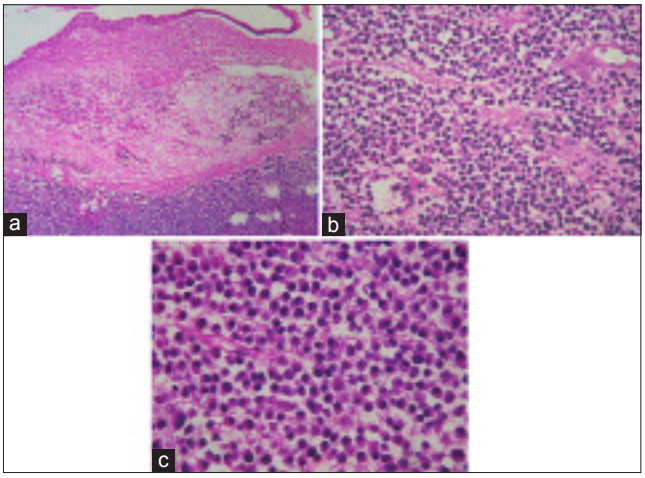

Figure 2: (a) Microscopy shows a neoplasm with overlying respiratory epithelium ×40, (b) sheets of plasma cells ×400 and (c) high power view of plasma cells ×1000. (a-c: H and E) with moderate amount of cytoplasm and eccentric round nuclei

Table 1: Summary of the reported cases of clival plasmacytoma including the present case

Authors	Age	Sex	Clinical features	Radiology findings	Diagnosed by	Treatment	Follow-up	Remark
Vera *et al.* 1980[6]	49 years	Male	Incidental	Destructive lesion	Biopsy	Radiation	14 months doing well	
Bindal *et al.* 1995[7]	43 years	Female	Headache	Not available			No recurrence	
	47 years	Male	Headache	Not available			Stable for 25 years	
Goyal *et al.* 2006[4]	60 years	Male	Headache, diplopia-2 months	Expansile lytic lesion	Biopsy	Not available		
Mahale *et al.* 2007	58 years	Male	Decreased hearing, earache-1-month	Destructive lesion with calcification	Cytology	Not available		
Liu *et al.*[7] 2010	54 years	Female	Hemianopia-40 days	Lytic lesion	Biopsy	Subtotal resection followed by radiation	22 months doing well	
Guinto-Balanzar *et al.*[3] 2012	66 years	Female	Headache, diplopia-3 months	Homogenously enhancing lesion	Biopsy	Complete resection	Died 3 months after surgery	
	61 years	Male	Headache tinnitus-6 months	Infiltrating lesion	Biopsy	Subtotal resection followed by chemo-radiation	3 years doing well	Associated with underlying MM
Liu and Qiu *et al.*[2] 2012	40 years	Male	Horizontal diplopia	Homogenously enhancing expansile lesion	Endoscopic biopsy	Radiation, followed by high dose steroids	4 months later, PET scan showed increased uptake, underwent gamma knife surgery	3 months doing well
Present case	55 years	Male	Diplopia	Expansile isointense lesion	Biopsy	Subtotal excision followed by radiation	8 months doing well	

MM – Multiple myeloma; PET – Positron emission tomography

the diagnosis is obtained through histopathology, as in our patient also.

Histologically, plasmacytomas are characterized by a diffuse or sheet-like proliferation of plasma cells with varying degrees of maturity and atypia. The nuclei are oval to round and eccentrically located with a dispersed ("clock-face") nuclear chromatin pattern and a clear or halo area. Neoplastic nature of plasma cells is established by proving the monoclonality.

This case is presented to highlight that though rare, plasmacytoma should be considered in the differential diagnosis of skull base lesions associated with early involvement of cranial nerves. All solitary plasmacytomas patients should be evaluated to rule out underlying MM and thoroughly followed up for any signs of evolution to MM.

Average survival of patients with MM is 3 years, whereas patients with a solitary lesion have a better prognosis. EMP progressing to MM is <30% with a 10-year disease-free period of 70%. In SBP, however the likelihood of progression to MM is >50% with a 10-year disease-free period of only 16%.[3, 6, 7]

References

1. Barnes L, Eveson JW, Reichart P, Sidransky D. Pathology and Genetics of Head and Neck Tumours. Lyon: IARC Press; 2005. p. 61-2.
2. Liu Y, Qiu J. Solitary intracranial plasmacytoma located in the clivus: A diagnostic and theraputic challenge. North Am J Med Sci 2012;5:232-5.
3. Guinto-Balanzar G, Abdo-Toro M, Aréchiga-Ramos N, Leal-Ortega R, Zepeda-Fernández E, Nambo-Lucio Mde J. Plasma cell tumor of the clivus: Report of two cases. Cir Cir 2012;80:171-6.
4. Goyal R, Gupta R, Radotra BD. Plasmacytoma of the clivus: A case report. Indian J Pathol Microbiol 2006;49:568-70.
5. Mahale A, Ullal S, Thiagarajan D, Das S. Plasmacytoma of the base of skull – A case report. Indian J Med Paediatr Oncol 2007;28:34-7.
6. Vera CL, Kempe LG, Powers JM. Plasmacytoma of the clivus presenting with an unusual combination of symptoms: Case report. J Neurosurg 1980;52:857-61.
7. Liu ZY, Qi XQ, Wu XJ, Luo C, Lu YC. Solitary intracranial plasmacytoma located in the spheno-clival region mimicking chordoma: A case report. J Int Med Res 2010;38:1868-75.

How to cite this article: Amita R, Sandhyamani S, Nair S, Kapilamoorthy TR. Plasmacytoma of the clivus. Asian J Neurosurg 2017;12:573-5.

Source of Support: Nil, **Conflict of Interest:** None declared.