Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2014; 2(4):308-310 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2014.v02i04.0029

Spinal Paraganglioma- A Rare Case Report

Dr ZahedaKausar[1*], Dr Syed ShafiqurRahman[2], Dr syedJaleelkirmani[3], Dr Jaipal Reddy[4], Dr Amanulla khan Mohmmed[5]

[1]Assistant professor upgraded department of pathology, Osmania General Hospital, Hyderabad, India,
[2]Consultant Radiation Oncologist, Bibi Cancer Hospital , Chanchalguda, Hyderabad, India
[3]Associate professor, Osmania General Hospital, Hyderabad, India,
[4]SK Neuropolytrauma Hospital, Hyderabad, India,
[5]Post Graduate, Upgraded department of pathology, Osmania General Hospital, Hyderabad, India,

***Corresponding Author:**
Name: Dr ZahedaKausar
Email: zahedar11@gmail.com

Abstract: Paragangliomas of the spine are rare neuroendocrine tumours. We present a case of Paraganglioma of the spine (L2-4), clinically diagnosed as lumbar schwannoma. A 40 year old female presented with low back pain for which she was investigated. Her MRI showed intradural / extra medullary well defined mass lesion at the L2-4 level. She underwent surgical excision. Histopathological examination of the resected tumour mass revealed features suggestive of paraganglioma. The tumour cells were positive for chromogranin, synaptophysin and S100 .Although Paragangliomas are rare in spinal region, and it should be included in differential diagnosis.
Keywords: chromogranin;Radiculopathy ; neuroendocrine; Spinal paraganglioma; Spinal tumour;zell –ballen

INTRODUCTION

Spinal paragangliomas are neuroendocrine tumours of the extra adrenal paraganglionicsystem.Carotid and glomusjugulare tumours constitute more than 90% of extra adrenal paragangliomas. Spinal paragangliomas are very rare [1]. The incidence of spinal paraganglioma is 0.07 per 100000 seen mostly in mid life (age range (13-71) with slight male preponderance. spinal paraganglioma was described by Miller and Torack in 1970. Commonly these patients present with low back pain and monoradicular symptoms because these tumours present as spinal mass lesion. Diagnosis is based on radiological imaging, histopathology .immunohistochemistry. The treatment of these tumours is excision with or without radiotherapy [2]. We are presenting this case because of its rarity.

CASE REPORT

A 40 years old female presented with history of low back pain for 1year .Her general, physical, and systemic examination was normal. Her routine investigations including ultrasound abdomen was nil remarkable, but magnetic resonance imaging of lumbar spine showed uniformly enhancing well circumscribed intradural lesion at L2-L4 level (fig 1). On imaging impression of schwannoma was made. She underwent L2-L4 laminectomy, durotomy and excision of lesion done. Patient had an uneventful postoperative period. Grossly we received grey brown tissue mass measuring 5x4 cm in size cut section showed solid grey brown areas. Microscopic examination of the hematoxylin – eosin staining revealed tumour cells arranged in zell-ballen pattern (fig 2). The individual cells had moderately abundant cytoplasm and centrally placed round to oval nuclei with salt and pepper type of chromatin. There were no mitotic figures. There was no necrosis so diagnosis of neuroendocrine tumour was made on immunohistochemistry tumour cells were positive for chromogranin, synaptophycin, and interspersed cells were positive for s-100(fig 3). With these features the diagnosis of paraganglioma was confirmed. The patient underwent post operative radiotherapy and the patient is on follow up.

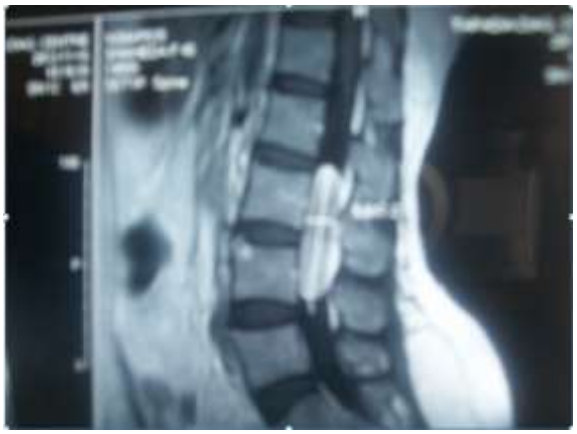

Fig-1: MRI lumbosacral spine sagital section shows isointenseintradural lesion at L3 level.

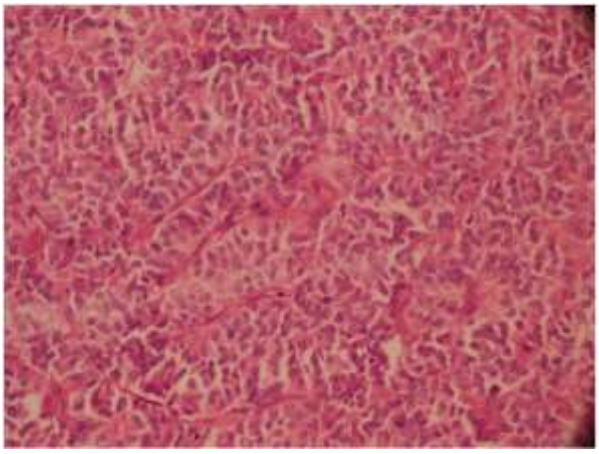

Fig-2: photograph showing tumour cells arranged in Zell-ballen pattern H&E X 40.

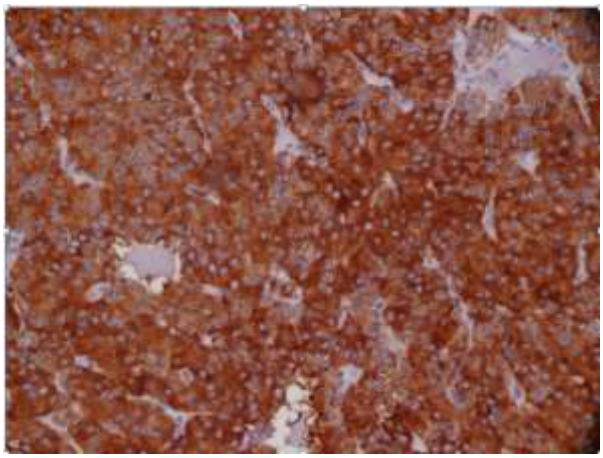

Fig-3: photograph showing diffuse positivity of tumour cells to chromogranin X 40

DISCUSSION

Paragangliomas are tumours derived from neural crest origin [3]. The most common sites of paraganglioma are adrenal medulla, carotid bodies and glomusjugulare which account for 80-90% of the cases . The unusual sites where paragangliomas are seen are pineal region, sellaturcica, suprasellar region or the orbit. Spinal location is rare. The first case of spinal paraganglioma was described by Miller and Torack who described it as a secretory ependymoma of filumterminale [4].The location of these tumours is intradural and extramedullary. They are most commonly seen in lumbar region as it is noted in our case followed by cervical and thoracic regions [5]. Spinal paragangliomas may originate from sympathetic neurons situated in thoracic and lateral horns of the spinal cord which send their axons to the sympathetic trunk through communicating branches.

Paragangliomas of caudaequina are seen mostly in midlife (age range 13-71yrs) with slight male preponderance [6].in our case the patient is female 40 yrs. These are present with signs and symptoms of spinal mass and may cause radiculopathy.the commonest nerve root involved is L3[7].our patient presented with low back ache magnetic resonance imaging show characteristic well defined lesion.

According to Lecompteparagangliomas are classified according to histological basis as organoid, angiomatous and adenomatous types(nayilkhurshid)spinal paraganglioma)microscopically these tumours show typical nuclear monomorphism and all display zell-ballen pattern(large polyhedral chief cells arranged in nests),many show granular argyrophilic and argentaffinreaction, areas of haemorrhage and necrosis may be seen most of these tumours have low proliferative potential MIB-1 labelling index usually ranges from 0.01-0.2%.malignancy cannot be determined with histological assessment but is demonstrated by metastasis and invasion, immunohistochemistry is used for conformation of diagnosis. Immunohistochemistry is positive for chromogranin, synaptophysin, neuron specific enolase and s100, all these markers showed positivity in our case.

The majority of paragangliomas of caudaequina are amenable to surgery and total excision should be done after excision prognosis is good. Only 4% of spinal paragangliomas recur after total excision. Radiotherapy is reserved for locally invasive tumours or where excision has been incomplete in our case radiotherapy was given because of incomplete excision. In the WHO grading system paragangliomas are classified as grade I [8].

CONCLUSION

Differential diagnosis of spinal intradural tumour, especially of lumbar area. These are benign neoplasms and need to be distinguished from more aggressive tumours as the prognosis after total excision is excellent.

REFERENCES

1. Soffer D, Pittaluga S, Caine Y, Feinsod M; Paraganglioma of caudaequine. A report of a case and review of the literature.Cancer, 1983; 51(10):1907-1910
2. Rumana M, Khursheed N, Vani S; Primary spinal paraganglioma :A review. Neurosurgery Quarterly 2009;19(3):196-199.
3. Boker DK, Wassmann H, Solymosi L; Paragangliomas of the spinal canal. SurgNeurol,1983;19(5): 461-468.
4. Miller CA, Torack RM; Secretory ependymoma of the filum terminale. Acta Neuropathol, 1970; 15(3): 240-250.
5. Rees JH, Smirniotopoulous JG, Moran C, Mena H; Paragangliomas of the caudaequina. Imaging

features with radiologicpathologic correlation. Int J Neuroradiol, 1996; 2: 242-250.

6. Sonneland PRL, Scheithauer BW, LeChago J, Crawford BG, Onofrio BM; Paraganglioma of the caudaequina region. Clinicopathological study of 31 cases with special reference to immunocytology and ultrastucture. Cancer, 1986; 58: 1720-1735.
7. Rumana M, Santosh V, Khursheed N, Yasha TC, Kolluri VR, Shetty S, Ravi KC; Primary spinalparagangliomas: a clinicopathological and immunohistochemical study of six cases. Indian J PatholMicrobiol, 2007; 50(3): 528-532.
8. Sundgren P, Annertz M, Englund E, Stromblad LG, Holtas S; Paragangliomas of spinal canal. Neuroradiology, 1999; 41(10): 788-794.