Spinal paraganglioma presenting as a lumbar canal stenosis – a rare case report

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ABSTRACT

Paragangliomas arise from para ganglion cells. These are slow growing tumors (< 2 cm in 5 years) and are histologically benign (< 10% associated with lymphnode involvement or distant spread). Most contain secretory granules on electron microscopy (mostly epinephrine & nor-epinephrine, and these tumors may occasionally secrete these catecholamines with risk of life-threatening hypertension and/or cardiac arrhythmias). Carotid bifurcation most common site. Spinal paragangliomas are very rare neuroendocrine tumors often presenting with low back pain and radicular symptoms. Radiographically spinal paragangliomas mimic more commonly described tumors, such as ependymomas, schwannomas, meningiomas, and even hemangiomas, but a "salt and pepper" appearance related to a serpiginous vascular structure is instructive. Indeed, the rarity of this tumor makes the diagnosis rather challenging radiographically. We report a case of 55yrs old female patient with clinical presentation like lumbar canal stenosis on evalution found intra dural spinal canal tumor at L3and L4 vertebra,suspected to be schwanomma radiologically. After excision of the specimen and sent for histopathological examination suggestive of ependymoma. After immunohistochemistry study of tumor found to be paraganglioma.

Keywords: Paraganglioma, Spinal Tumour, Intradural Tumour

INTRODUCTION

Paragangliomas are originating from the autonomic nervous system, and are found in adrenal and extra adrenal locations. Extra adrenal paragangliomas are rare and occur most commonly in carotid bodies and the jugular glomus

Primary spinal paragangliomas are extremely rare and most frequently involving caudaequine and filumterminale[1,2,3]

CASE REPORT

55 year old female, presented to neurosurgery outpatient department with complaints of low back pain since 4years.Pain was increasing since 6 months and was not responding to any kind of treatment. Pain was radiating to both lower limbs more on to the right than left. She was not able to walk for long distance as she was getting paresthesias of both limbs.She had multiple acute episodes for the last six months; during the episode she was totally bed ridden and unable to fulfil her activities of daily living. Other than back pain, she had urinary hesitancy and constipation due to severe back pain. There was no weakness. When we examined the patient power in both lower limbs was 5/5 with 2+ reflexes. Gait was normal, antalgic type during pain. With the history and examination our provisional diagnosis is degenerative disc disease - probable -Lumbar canal stenosis. Patient visited multiple clinics and hospitals over a period of 4 years and finally presented to neurosurgery out patient department. On complete evaluation with chronicity of the symptoms, we advised magnetic resonance imaging (MRI) of lumbo sacral spine with whole spine screening

Magnetic resonance imaging showed as in image 1, image 2 and image 3.

Image 1 shows T2WI¬-- hyper intense lesion (sagittal cuts) and T1WI-iso intense lesion at L3-4 level (sagittal cuts) seen while in image 2 myeogram showed lesion and image 3 was on contrast study homogenous enhancement of the lesion was seen.

Image 1

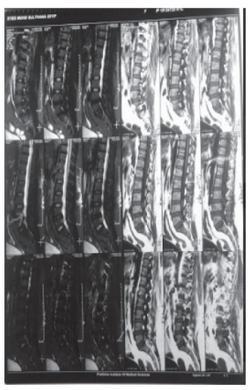


Image 2

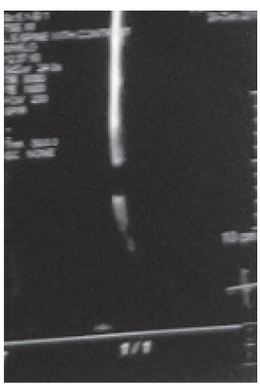
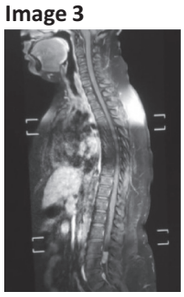


Image 3



Radiological diagnosis Possibilities were Schwannoma, Neurofibroma, Filum terminale Ependymoma And least possibility Meningioma

Patient was planned for surgery.Resection for the purposes of decompression of her nerve roots as well as to obtain pathology sample . After signing informed consent, she underwent resection.The patient underwent standard anesthetic induction and intubation, followed by prone positioning. The intended L3-4 space was localized using fluoroscopy and a subperiosteal dissection was performed, followed by an L3-4 laminectomy done.Durotomy and tumor excision was performed with thehelp of microscope

Operative findings were tumor was reddish pink in color,lobulated(image 4) and was not adherent to any roots .Tumor was attached to the filum terminale. Total resection of the tumor done and sent for histopathological examination.a dural closure was performed after meticulous intradural hemostasis was obtained. Hemostasis was then achieved in the surgical cavity and copious antibiotic irrigation was applied. The muscle, fascia, subdermal, and dermal layers were subsequently closed in a standard fashion.The patient was then turned to the supine position and extubated. Her immediate postoperative examination demonstrated a stable preoperative examination, including 5/5 strength in her bilateral lower extremities.

Figure 4-intra op picture

Figure 5-excised tumor specimen

Image 4

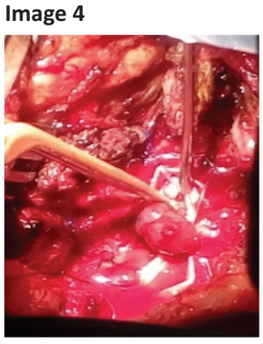
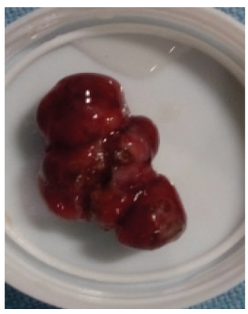


Image 4



Pathology

Histological sections Composed of columnar cells arranged around a central lumen i.e.ependymal rosettes

Suggestive of ependymoma as shown in image 6 &7. We advised immunohistochemistry of the sample and

Image 5

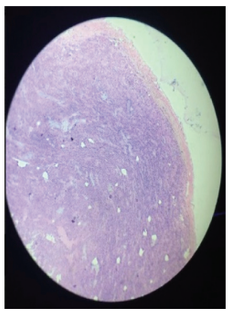
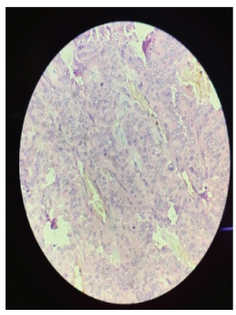
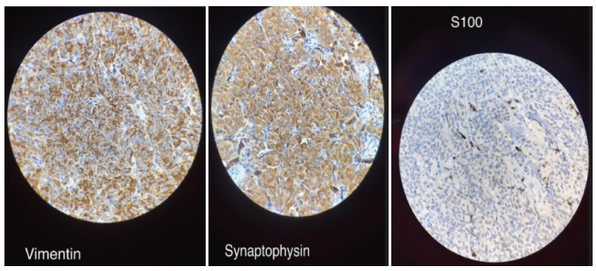


Image 6



Immune histochemistry showed vimentin&synaptophysin positive and s100 shows sustenticular cells. These features were suggestive of paraganglionoma as in image 8

Image 8



DISCUSSION

Paragangliomas are classified as World Health Organization (WHO) Grade I given their characteristically slow growth and histologically benign appearance. As they are neurogenic and arising from the autonomic nervous system, they are

Sub-classified into sympathetic – secreting catecholamine or parasympathetic and thus functionally non-secretory In the central nervous system, 80-90% of paragangliomas are identified in at the bifurcation of the common carotid artery and in the middle ear and predominantly of the parasympathetic type. Spinal paragangliomas are even less common and generally occur at the lumbosacral extent of the spine without reported variation in distribution. Male to female ratio of nearly 1.7:1 has been reported. The average duration of symptoms preceding diagnosis is typically around two years on the short end of the interval. Low back pain and sciatica were the most common clinical presentations, and sensory-motor and sphincter dysfunction can develop along with clinical progression. Some cases can present with symptoms of radiculopathy or slowly progressive spinalcord compression[4]

Preoperative diagnosis is determined using magnetic resonance imaging, but due to the rarity of paragangliomas,

radiographic diagnosis is challenging.On magnetic resonance imaging (MRI), paragangliomas are generally hypo intense to isointense on T1WI and isointense to hyper intense on T2WI; T1WI homogeneity in contrast enhancement helps to delineate the tumor.A characteristic "salt & pepper" appearance on T2WI is often described resulting from a rich vascular nature of paragangliomas; serpiginous flowvoids are often evident on T2WI and occasionally peripheral hemosiderin is evident as a hypo intensity in the T2WI.Radiographic differential diagnoses include myxopapillaryependymoma, schwannoma, meningioma, teratoma, and hemangioma[5]

Surgical pathology provides the definitive diagnosis of resected lesions A "Zellballen" pattern as a solid mass of the paraganglion-type nests or cords is often described. Immunohistochemical staining is often S-100 positive while Ki-67 proliferative markers are generally low, suggesting a benign tumour of neuronal origin.Long-term follow-up data for management of spinal paragangliomas is sparse. In the largest analysis of spinal paragangliomas, Xu and colleagues followed 19 cases and determined the mean age of presentation to be 47.7years. Initial presenting symptoms typically included back pain with sciatica ,which immediately resolved with operative resection; three of five cases with incomplete resection showed recurrence of disease at 62.1 months.[6]

Very few cases have been reported in literature[7,8,9,10] and hence more and more reporting will help in better understanding of the condition.

Gross total resection is advocated but sometimes found difficult due to adhesion to functional nerve roots. Data for adjunct treatment modalities also remains sparse. Conventional radiotherapy affords control rates of 90-100% for intracranial paragangliomas and is often advocated as a first-line therapy for asymptomatic patients with intracranial paragangliomas.

CONCLUSION

Spinalparagangliomas are a very rare and often presenting with low back pain and radiculopathic symptoms.

clinical course of the disease is benign with operative resection as the first-line treatment with nearly immediateimprovement in symptoms.Radiographic characteristics include a vascular, "salt and pepper" appearance with prodigious flow voids and an

Occasional hemosiderin rim.Gross total resection.The role of adjunctive radiotherapy is unknown but, borrowing from the intracranial paraganglioma literature, maybe useful in long-term progression management.

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