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RETROPERITONEAL PARAGANGLIOMA - A CASE REPORT

NALLI. R. SUMITRA DEVI¹, N. SANGEETHA², N. HEMAVATHY³, C. ARUNPRABHAKARAN⁴

1. Professor, Department of pathology, Dr. M. G. R Tamil Nadu Medical University, Stanley Medical College, Chennai, Tamil Nadu, India.
2. Assistant Professor, Department of pathology, Dr. M. G. R Tamil Nadu Medical University, Stanley Medical College, Chennai, Tamil Nadu, India.
3. Assistant Professor, Department of pathology, Dr. M. G. R Tamil Nadu Medical University, Stanley Medical College, Chennai, Tamil Nadu, India
4. Postgraduate, Department of pathology, Dr. M. G. R. Tamil Nadu Medical University, Stanley Medical College, Chennai, Tamil Nadu, India

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Abstract: Introduction-Retroperitoneal paragangliomas are rare endocrine tumors and are easily misdiagnosed. These tumors are often discovered incidentally during imaging studies performed for other reasons. Paragangliomas are tumors arising from extraadrenal medullary neural crest derivatives. They are usually located in the head and neck, chest cavity, abdomen, bladder and pelvis. We report a case of paraganglioma of the sympathetic ganglia who presented with hypertension. Case report: 29 years old male came with the complaint of abdominal pain for 3 months duration. On examination no abdominal mass was palpable and his BP was 150/100 mmHg. Urine catecholamines were normal. Surgical excision of the mass was performed. Histopathology revealed paraganglioma of the sympathetic ganglia. Conclusion: We present a case of paraganglioma from the sympathetic ganglia in the retroperitoneum for its rarity. Definitive diagnosis is usually made with histological findings and the treatment of choice is surgical resection followed by prolonged follow up.

Keywords: Histopathology, paraganglioma, Retroperitonem, sympathetic ganglia.



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Corresponding Author: MS. N. SANGEETHA

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INTRODUCTION

Retroperitoneal paragangliomas are rare tumours accounting for 10-15% of all paragangliomas with the incidence rate of 2-8 cases/million per year [1], and present several therapeutic challenges because of their rarity and relatively late presentation at the time of diagnosis. Paragangliomas arise from primitive cells of the neural crest [2] that during embryogenesis migrate to several sites in the body [3]. The primitive cells differentiate into paraganglionic chief cells and supportive sustentacular cells forming autonomic nerve clusters designated paraganglia. Those in the retro peritoneum and thorax are associated with sympathetic nerves and in head and neck with parasympathetic nerves [4, 5]. Second in frequency to those in the head and neck are intra abdominal paragangliomas, which arise in the organ of Zuckerkandl in the retroperitoneum [6]. They may be found at any level in the retro peritoneum but usually are seen in the paraaortic sites. Functional paragangliomas secrete nor epinephrine and cause hypertension [6].

CASE REPORT

29 years old male came with the complaint of abdominal pain for three months duration. He had no symptoms related to catecholamine excess such as palpitation, headache and diaphoresis. On examination, no abdominal mass was palpable. He had a history of hypertension and his BP was 150/100 mmHg at the time of admission. Abdominal CT showed a multiloculated vascular cystic lesion between the aorta and inferior vena cava, lifting the pancreas. Urine VMA level was normal. Surgical excision of the mass was performed and sent for histopathological examination. Grossly we received multiple irregular friable greybrown soft tissue fragments largest measuring 7x6x1 cm and smallest measuring 2x1x1 cm. Cut surface appeared greybrown with focal hemorrhagic areas [Figure 1]. Histopathology showed a neoplasm composed of tumor cells arranged in nests and clusters surrounded by delicate capillary network (Zellballen pattern) [Figure 2]. Cells were round to polygonal with finely granular eosinophilic cytoplasm and round to oval nuclei with finely stippled chromatin [Figure 3]. Focal area showed a nodular aggregate of ganglion cells [Figure 4] with adjacent foci showing necrosis.



Figure 1 shows multiple grey-brown soft tissue fragments.

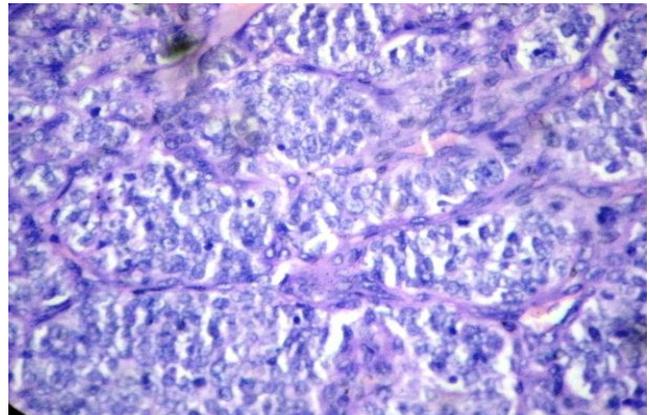


Figure 2 shows tumor cells arranged in lobules separated by vascular channels (zellballen pattern).

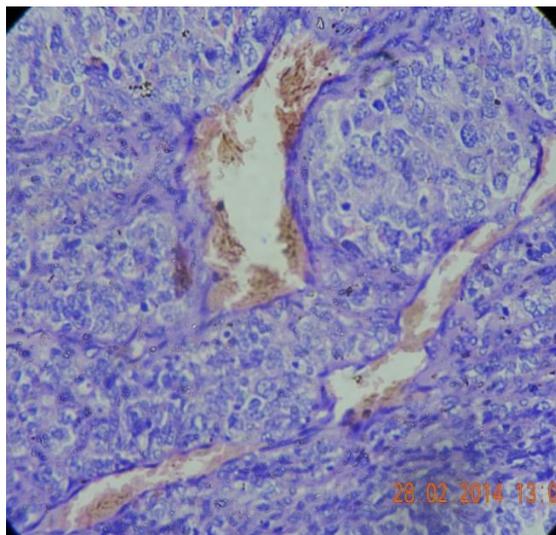


Figure 3 shows nucleus with finely granular chromatin and abundant eosinophilic cytoplasm.

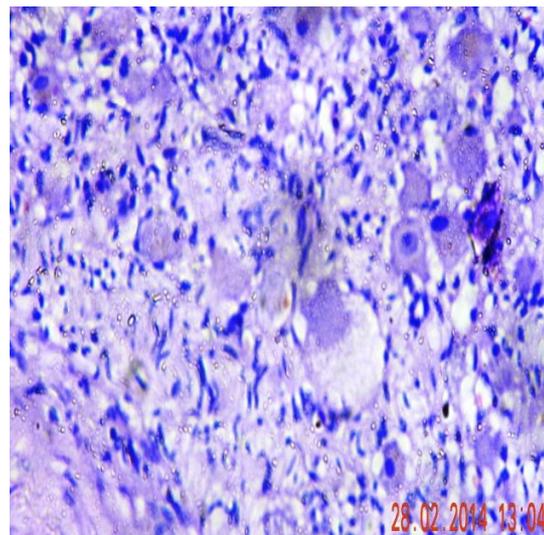


Figure 4 shows collection of ganglion cells.

DISCUSSION

The 2004 WHO classification of endocrine tumors, defines pheochromocytoma as a tumor arising from chromaffin cells in the adrenal medulla. Closely related tumors in the extra adrenal sympathetic and parasympathetic paraganglia are classified as extra adrenal paragangliomas [7]. Paragangliomas can be found from the upper cervical region to the pelvis along the autonomic nervous system. They are most commonly found in the organ of Zuckerkandl at the aortic bifurcation. Most retroperitoneal paragangliomas occur in patients of 30 -50 years of age,

although the malignant forms are seen in the younger age group [8]. Genetic disorders involving mutations within the SDH D & B subunits and the VHG gene place an increased risk in the development of extra adrenal paragangliomas and adrenal pheochromocytomas respectively [9]. Patients with non-functioning retroperitoneal paragangliomas are usually asymptomatic until the tumor reaches the sufficient size to produce compressive symptoms on adjacent structures [10, 11]. Patients with functional paragangliomas have paroxysmal episodes of hypertension, palpitation, headache and profuse sweating.

Grossly, paragangliomas are circumscribed, firm red brown, often focally hemorrhagic. Necrosis and cystic change can occur. Histopathologically the tumor is arranged in Zellballen pattern showing cell nests surrounded by a network of capillary channels with foci showing broad anastomosing and trabecular patterns. The neoplastic cells are oval to polyhedral contain a pale eosinophilic granular cytoplasm and round to oval vesicular nuclei with stippled chromatin rarely cytoplasm can be oncocytic [12]. The cytoplasm of functional catecholamine secreting paragangliomas is more abundant and grey in H&E sections. There may be accentuation of the nesting or alveolar pattern due to congestion with separation of nests of neoplastic cells. A variety of stromal alterations such as old or recent hemorrhages, fibrosis and vascular sclerosis can be present in some cases. Stromal changes, such as prominent sclerosis may cause difficulty in diagnosis but usually some parts of the tumor have a well preserved organoid pattern [13]. Cellular pleomorphism and hyperchromasia are not reliable features for a diagnosis of malignancy. Determination of malignant behaviour is based on the presence of metastasis to regional lymph nodes and to distant sites such as lung, liver and bones [5]. Confluent necrosis, mitotic figures and vascular invasion are seen more often in malignant than in benign paragangliomas. Sustentacular cells were found to be reduced in number in locally invasive tumors and rare or absent in malignant tumors.

4. CONCLUSION

Our case emphasizes the necessity to include extra adrenal paraganglioma in the differential diagnosis of retroperitoneal tumors, despite its rarity, yet the gold standard of diagnosis remains a pathological one. Lifelong follow up of patients with retroperitoneal paragangliomas is essential as metastasis and recurrences may occur.

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