

Paraganglioma: A diagnostic dilemma

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Abstract

We report a case of intra abdominal paraganglioma in a 46 year old male, that measured 12.5x10x10 cm and weighed 340 grams. A CT scan was done for the evaluation of an abdominal mass. The tumour was surgically removed and followed by an uneventful post operative recovery. On follow up at one year patient had no active complaints. Histologically paraganglioma was composed of tumour cells that had abundant, granular to clear eosinophilic cytoplasm and mildly pleomorphic, vesicular rounded nuclei. Occasional mitoses was seen. No atypia was noticed. Immunohistochemistry confirmed the presence of chromogranin, synaptophysin and vimentin markers. These are diagnostic for paraganglioma.

Introduction

Collections of neuroendocrine cells dispersed throughout the body are known as paraganglia and the tumour arising from these paraganglia are known as paragangliomas. Some paraganglia are connected with the sympathetic nervous system whilst others are connected with the parasympathetic nervous system. Paragangliomas are rare tumours, majority of which are benign and cured by surgical removal.

The paraganglia based on their anatomic distribution can be divided into three groups: (1) branchiomeric (2) intravagal and (3) aorticosympathetic. The branchiomeric and intravagal paraganglia are associated with the parasympathetic nervous system and are located close to the major arteries and cranial nerves of the head and neck. The aorticosympathetic paraganglia, as the name suggests, are located alongside of the abdominal aorta and are associated with the sympathetic nervous system. The paraganglia comprise chromaffin cells. They secrete and store catecholamines. This system is vital in foetal development until the formation of the adrenal medulla. Some of these paraganglia sense oxygen and carbon dioxide tensions within adjacent vessels, hence the tumours at times are known as chemodectomas.¹

Case Report

A 46 year old male, smoker for ten years with no comorbidities, presented with a painless abdominal mass for the last eighteen months. There was no history of weight loss, fever or any other associated symptom. Examination revealed an intra abdominal, non tender mass, extending from the level of the umbilicus into the pelvis (Figure 1A and 1B). The lower margin was impalpable and there was minimal

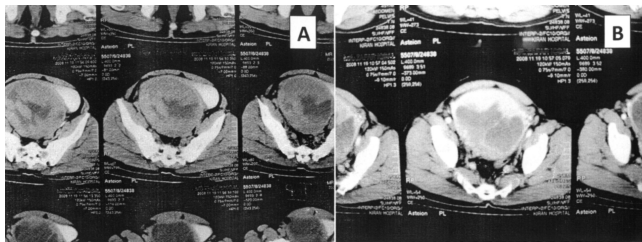


Figure-1 A & B: C.T abdomen with contrast: axial image showing the abdominal mass, abutting the surrounding structures.

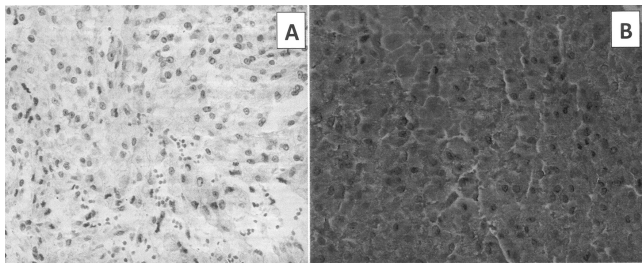


Figure-2 A & B: Immunohistochemistry results showing presence of S-100 (Figure 2A) and chromogranin (Figure 2B) markers.

mobility in the horizontal or the vertical axis. There were no signs of inflammation and it was firm to hard in consistency.

Patient complaints were pressure related symptoms and with a view of excision of the tumour or debulking it, a laparotomy was planned. A CT abdomen was done as a part of the workup to evaluate the tumour extension. The finding of the scan showed a tumour 11x14cm in its maximum dimensions, it was well defined marginally enhancing heterogeneous mass in the lower abdomen. This mass was hypo dense in the centre representing necrosis. It appeared to be abutting the urinary bladder with intact fat plane. Posteriorly it was also abutting the ureters and the pelvic vessel with intact fat planes. The remaining routine investigations were normal. The operative finding revealed an encapsulated mass lying just below the abdominal wall. The tumour was approximately 12.5 cm in maximum diameter displacing the small intestine towards upper left side. Anteriorly mass was free however, posteriorly it was attached to the sacrum through its capsule wall. Also, at the level of T 12 it was adherent to the right ureters, to which there was an iatrogenic injury (repaired at the same time). The tumour did not invade any other major structure and was removed in toto. Post operatively patient did well and went home on the fourth day.

The histopathology gave a gross examination revealing a brownish circumscribed mass with a partly ruptured capsule. Cut surface showed gray white friable masses, haemorrhagic brown areas and dilated blood vessels.

Microscopically it comprised nests and trabeculae of polygonal cells enclosed by thin walled vasculature and fibrous septae with few sustentacular cells. The tumour cells

had abundant, mildly pleomorphic rounded nuclei. Occasional mitoses / 10 hpf were seen. Large foci of necrosis and hyaline changes were seen. No atypia was noticed.

Immunohistochemistry results revealed that Chromogranin, Synaptophysin and Vimentin markers were diffusely positive in tumour cells along with S-100 which were negative in tumour cells but focally positive in sustentacular cells (Figure 2A and 2B). The following markers CK AE1/AE3, CD 117, HMB-45 and CD 34 were negative in tumour cells. Although CD 34 markers were found to be positive in the blood vessel wall.

Discussion

Literature tells us that rarely paragangliomas have also been reported in unusual sites such as the gall bladder,² mesentery,³ kidney,⁴ prostate⁵ and ovary.⁶ Paragangliomas maybe hereditary and can be associated with familial paraganglioma, neurofibromatosis type I, Von Hippel-Lindau disease, the Carney triad, multiple endocrine neoplasia type II and mutations of the succinate dehydrogenase gene (SDHB, SDHC and SDHD).

Predictors of malignancy in a paraganglioma include extra-adrenal location, confluent tumour necrosis, vascular invasion, local invasion, coarse nodularity and absence of hyaline globules.⁷ Though there is a documented evidence of malignancy in most tumours, in our case, the above described tumour was benign.

Majority of the patients with extra-adrenal abdominal paraganglioma presented with complaints of abdominal pain and/or a palpable abdominal mass,⁸ however patients may be asymptomatic. Nausea, vomiting, diarrhoea, abdominal distention and weight loss are the other symptoms reported by patients with paraganglioma. Symptoms such as hypertension, flushing, sweating, headache, diaphoresis, anxiety, tachycardia or palpitations are symptoms reported in patients with increased catecholamine secretion in functional tumours.⁸⁻¹⁰

Even though imaging techniques are helpful, diagnoses of extra abdominal or intra abdominal paragangliomas can only be done with careful histological and immunohistochemical evaluation. CT scan is useful for diagnosis while MRI has the highest sensitivity in detection of extra-adrenal paragangliomas as well as pheochromocytomas.^{11,12}

The treatment of choice for a paraganglioma is complete surgical resection followed by a prolonged follow up. This may be done by six monthly ultrasound or yearly CT scans. There is no specific recommended follow up strategy.

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