MANAGEMENT DILEMMAS OF RETROPERITONEAL PARAGANGLIOMA: REPORT OF A CASE

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MANAGEMENT DILEMMAS OF RETROPERITONEAL PARAGANGLIOMA: REPORT OF A CASE (Abstract): A 17-year-old boy presented to emergency department with abdominal pain in the left upper quadrant along with a palpable mass. A CT scan revealed a 10 x 8.5 cm mass in the retroperitoneal area, suggestive of a sarcoma. The patient underwent surgical resection of the lesion, the histological examination of which confirmed a paraganglioma. An ¹³¹I-Metaiodobenzylguanidine (MIBG) scan excluded distant metastasis. Paragangliomas are rare tumours of the neuroendocrine system and certain considerations are pivotal in the management of these tumours. Urinary and plasma catecholamines measurement along with CT, MRI, and MIBG scanning facilitate diagnosis, localisation and characterisation of these tumours. Surgical excision is the treatment of choice for both primary and recurrent tumours. Unfortunately, histological characterisation for malignancy is limited and these tumours are considered malignant upon development of metastasis or local recurrence. Inexistence of histological criteria for malignancy means a long-term surveillance is the norm in these patients. Keywords: ADOLESCENT, PARAGANGLIOMA, RETROPERITONEAL

Paragangliomas are the rare tumours arising from the extra-adrenal neuroendocrine cells (1). Most paragangliomas occur in head and neck region and occurrence at other sites including the retroperitoneal area is relatively uncommon (1,2). Although a familial predisposition has been described, these tumours are seldom considered in healthy teenagers (3). We report a case of an adolescent boy who presented with severe abdominal pain and a palpable mass. The diagnosis was established on histological examination of the tumour. This report is aimed at discussing certain diagnostic and management challenges, and providing the clinicians with a concise account of how to address those.

CASE REPORT
A previously healthy 17-year-old boy attended the emergency department with severe abdominal pain of a day’s duration. A physical examination revealed a palpable mass in the left upper quadrant of the abdomen. The patient was apyrexial and full blood count, urea and electrolytes
were unremarkable. On a CT scan, the mass appeared to be a heterogeneous tumour in the retroperitoneal area in close proximity to pancreatic tail and descending colon, measuring 10 x 8.5 cm. The tumour appeared to be a retroperitoneal sarcoma containing areas of calcification and with internal haemorrhage. Angiographic embolisation of the rich vascular arcade derived from the inferior mesenteric, subcostal, and lumbar arteries was achieved preoperatively to minimise the blood loss. At operation, the mass was found to be an encapsulated, highly vascular tumour occupying the left half of retroperitoneal area and adherent to the upper descending colon (fig. 1). Complete removal of the tumour was performed along with segmental resection of the involved descending colon. The histological examination of the tumour showed the features consistent with a paraganglioma.

The patient made an un-eventful recovery in the postoperative period and was discharged home on the 5th day after operation. A subsequent 131I- Metaiodobenzylguanidine (MIBG) scan in the early postoperative period excluded distant metastasis. Clinically, the tumour was considered benign in view of the non-metastatic disease, as discussed below. Nevertheless, the patient will require a long clinical follow-up due to inexistence of definable histological criteria for malignancy in paragangliomas.

DISCUSSION
The paragangliomas arise from the neural crest cells lying outside adrenal medulla. These tumours originate both in the hereditary as well as the sporadic form and are most commonly diagnosed in the fourth or fifth decade of life (3, 4). Anatomically, these tumours occur anywhere from upper neck to pelvic floor, however, head and neck remain the commonest regions affected (4, 5). Retroperitoneal paragangliomas usually develop in sympathetic paraganglia and at least half of these secrete catecholamines. Such tumours are called functional paragangliomas. It has been reported that up to half of the abdominal paragangliomas are malignant (5). The common presenting features include abdominal mass, pain, palpitations, headache, sweating, hyperten-
sion, and genitourinary symptoms (2, 5, 6).

The diagnosis is established by measurement of the urinary and plasma catecholamines, radiological findings of tumour on CT or MRI scan, and $^{123}\text{I}$- or $^{131}\text{I}$-MIBG scanning for functional paragangliomas (5, 7). Tumour localisation has improved remarkably owing to the use of different imaging modalities. Unfortunately, the histological examination is limited by its inability to distinguish between benign and malignant tumours, and tumour recurrence or metastasis are generally accepted as features of the malignant paragangliomas (8, 9). Therefore, these patients usually require a long clinical follow-up. Annual biochemical testing, CT or MRI scan, and MIBG scintigraphy are essential in the follow-up process to diagnose recurrence, delayed appearance of multiple tumours, or metastasis (4, 7, 8).

Surgical excision remains the mainstay of treatment for both primary and recurrent tumours (1, 4, 5, 7). Feasibility of the laparoscopic approach has been reported albeit open resection remains the commonest operative strategy (4, 10). The functional tumours require reversal of circulating catecholamines prior to the operation. Both alpha and beta antagonistic drugs are used preoperatively to control blood pressure and to prevent intraoperative hypertensive crisis (4, 7). Radiotherapy or chemotherapy may be used for preoperative debulking of tumour. Ablation of the tumour with high dose I-131 MIBG has shown promising results and more evidence is awaited (5, 11).

The abdominal paraganglioma is an uncommon condition in adolescents (6, 12). Therefore, it remains a rare clinical consideration in previously healthy teenagers presenting with abdominal pain. In managing such cases, a degree of suspicion should be maintained for uncommon pathologies like neuroendocrine tumours. A careful clinical assessment cannot be over emphasised which would help detect an associated mass. Absence of acute inflammatory markers should trigger instigation of further investigations, the most appropriate being a CT or an MRI scan. It is vital that such cases are assessed and managed on an individual basis given the implications of misdiagnosis.

CONCLUSIONS

Extra-adrenal retroperitoneal paraganglioma presents diagnostic and management challenges and, once suspected, certain management considerations are pivotal. A careful clinical assessment differentiates it from common causes of pain in this age group. The urinary and plasma catecholamines measurements along with CT, MRI, and MIBG scanning should be employed to diagnose, localise and characterise these tumours. Surgical excision is the treatment of choice for both the primary and the recurrent tumours. Inexistence of histological criteria for malignancy means a lifelong surveillance is the norm in these patients.

REFERENCES


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**NEW APPROACH IN TREATMENT OF MULTIDRUG-RESISTANT TUBERCULOSIS**

Multidrug-resistant tuberculosis (MDR-TB) is a major problem health concern. According to the World Health Organization (WHO), in 2012, MDR-TB accounted for 3.6% of new cases and 20% of previously treated cases of TB (1). Therapy for MDR-TB is long, hard and expensive with antiquated drugs. After 40 years, a new antituberculosis drug – bedaquiline - has become available for use. Bedaquiline is an inhibitor of mycobacterial ATP synthase with bactericidal activity. A recent clinical trial, which included 160 new cases of MDR-TB treated with bedaquiline in combination with background treatment, showed that the patients who received bedaquiline have had a median time to sputum-culture conversion by 83 days, with 42 days shorter than placebo group. Also, bedaquiline used in therapeutic regimen for MDR-TB determined an increase of rate of culture conversion at 24 weeks (79%) and at 120 weeks (62%) compared with placebo (58% respectively 44%). The same study showed that the risk of acquiring new resistance to anti-TB drugs was lower in patients treated with bedaquiline (2). These findings support the use of bedaquiline in MDR-TB treatment (World Health Organization. Global tuberculosis report 2013. www.who.int/tb/publications/global_report/en/; Diacon AH, Pym A, Grobusch MP et al. Multidrug-resistant tuberculosis and culture conversion with bedaquiline. *N Engl J Med.* 2014;371:689-691, 723-732).