Mass in the Adrenal Region: Not Always of Adrenal Origin: A Diagnostic Dilemma.

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INTRODUCTION

Adrenal mass is an uncommon entity for which patients are referred to surgeons. The incidence of adrenal masses found on abdominal Computed Tomography (CT) scans is between 0.6% and 1.3%. The incidence of these masses on all CT scans, including thoracic, abdominal, and pelvic, is between 0.4% and 4.4%.¹,² The differential diagnosis of such masses are wide ranging from incidentalomas to adrenal cortical carcinomas. However it is very unusual for a posterior mediastinal mass to masquerade and present as an adrenal mass. Neurogenic tumors are the most common cause of a posterior mediastinal mass. They constitute 23% of all mediastinal masses in adults and generally are asymptomatic.³ However often they have rare presentations that make diagnosis and treatment extremely difficult. Benign schwannomas are the most common type of posterior mediastinal tumors. Often resection is enough and they tend to have excellent prognosis.⁴,⁵

CASE REPORT

A 55 yr old lady presented with the chief complaint of retrosternal chest pain since last two months. The pain was insidious in onset, dull aching in nature, mild in intensity and was not associated with palpitation, exertional dyspnoea, sweating or any other constitutional symptoms like cough, fever, headache, hemoptysis or recent loss of weight. There was no specific aggravating or relieving factor. However the pain later on migrated to the epigastric region from its initial site. She had two episodes of non bilious vomiting prior to hospitalisation. Initially she visited a general physician who ordered a Chest X Ray due to the nature of pain which showed a homogenous opacity in the lower part of right lung field. She was then referred to our hospital for surgical management. Detailed history was taken which was unremarkable. Her physical examination was also insignificant except slight tenderness in her epigastrium and a vague palpable mass in her right upper quadrant. USG showed a right adrenal heterogenous mass with mild heterogenous enhancement with irregular margins. Usg showed a right adrenal heterogenous mass with mild heterogenous enhancement with irregular margins. CT scan of chest and abdomen with contrast revealed following findings: "A large (95×63×78 mm) well defined soft tissue mass at right suprarenal region (SOL). Contrast enhanced CT scan revealed following findings: “A large (95×63×78 mm) well defined soft tissue mass at right suprarenal region extending into posterior mediastinum, with mild heterogeneous enhancement with irregular central non enhancing area but without calcification [Figure 1, 2]. The mass is compressing the Inferior Vena cava (IVC). The right adrenal gland not seen separately while the left one is normal in size and location”. However a CT Angiography done did not reveal any such vascular compression [Figure 3]. Her biochemical parameters including serum cortisol and urinary VMA was surprisingly normal. A Cardiothoracic consultation was made and the patient was posted for surgery in Vascular OT, keeping in mind a provisional diagnosis of non functioning adrenal tumour probably with mediastinal invasion. A right posterolateral thoracotomy was done through the 9th intercostal space under general anaesthesia with endotracheal intubation. Astonishingly the diaphragm could be palpated beneath the mass suggesting it to be a supradiaphragmatic tumor which

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Figure 1: Showing the heterogenous mass in the right adrenal area.

Figure 2: CT Thorax showing posterior mediastinal extension of the right adrenal mass.

Figure 3: CT Angiography showing the infra and suprarenal vessels.
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could be easily dissected from the surrounding pleura. The thoracotomy was closed and a right intercostal drain was placed. Postoperatively patient was managed with routine postoperative care. The patient made a speedy recovery and the patient was discharged on 5th postoperative day. The pathology report revealed the mass to be a benign schwannoma. On 10 months follow up she continues to be in excellent state of health.

DISCUSSION

Schwannomas are benign tumours which usually arise in the base of the spinal nerves, but sometimes may involve the thoracic nerves. They are often isolated lesions (as was in our case), however, multiple lesions along a nerve have been described in patients with Von Recklinghausen’s neurofibromatosis. A solitary neurofibroma rarely tends to be malignant, but in patients with Von Recklinghausen’s disease, 4% to 10% of the tumors become malignant.[6] Macroscopically, schwannomas are encapsulated, heterogeneous tumors with cystic degeneration.[4] It affects males and females equally, predominantly in the third or fourth decades of life.[7,10] Most patients are asymptomatic, although few may experience pain due to compression of the adjacent structures or intraspinal invasion of the tumor. Radiologically, they appear as spherical paraspinal masses with clean lobular margins, involve one or two posterior intercostal spaces, and can grow to large dimensions.[4,11] In 50% of cases, they cause benign erosions and deformities of the ribs, vertebral bodies, and nerve foramina. Routine chest radiographs rarely show calcifications.[4,10] However none of these standard descriptions fit with the radiological characteristics of the mass seen in our case. Schwannomas arise from the schwann cells of the nerve sheath. Histopathological examination remains cornerstone to diagnosis. Microscopically, they consists of spindle cells that give rise to dense cellular areas (Antoni A) and hypocellular areas (Antoni B).[7,9] Mitotic figures are rare. An adrenal mass is usually resected by either open or laparoscopic adrenalectomy while the standard approach for mediastinal neurogenic tumours is via posterolateral thoracotomy. The intraoperative discovery of a presumed adrenal mass (operated transabdominally) to be supradiaphragmatic poses tremendous challenge for a surgeon less versed with thoracotomy procedure. Though transdiaphragmatic resections of such supradiaphragmatic neurogenic tumors has been described in literature,[12] such expertise may not be available at all time. We advise to exert caution in adrenal masses with posterior mediastinal extension and explore them in liason with vascular teams. It would be wise to explore such indeterminate masses via the thoracic route before attempting abdominal exploration as the latter may prove futile.

CONCLUSION

This is probably the first case of a schwannoma, mimicking as adrenal tumor, being reported from India. This case highlights the importance of considering posterior mediastinal tumors in the differential diagnosis of adrenal masses and outlines the importance of clinical decision making in managing such ambiguous cases. Exploring the tumor by thoracotomy proved useful in our case and the patient was spared of unnecessary laparotomy and its morbidity.

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CONFLICT OF INTEREST

None

ABBREVIATION USED

VMA: Vanillyl mandelic acid; OT: Operation theatre

REFERENCES


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