

Pheochromocytoma: An Imaging Kaleidoscope

Dr. Priyanka Jain¹, Dr. Sahil Singla², Dr. Poonam Tanwar¹, Dr. Rohtas Yadav¹

¹Department Of Radiodiagnosis

²Department Of Surgery University Of Health Sciences Rohtak, India

Abstract: Pheochromocytomas are rare catecholamine secreting tumors of adrenal gland with 10% of cases found in children. They have a varied clinico-imaging spectrum of appearances and they may mimic other adrenal lesions, both benign and malignant. On Computed tomography, they may also demonstrate rapid contrast material washout and be mistaken for an adenoma owing to their deenhancement profile; however, their washout pattern can be inconsistent. Other atypical features include fatty, hemorrhagic, cystic, and calcific changes. Their early diagnosis is imperative due to poor clinical outcome and surgical complications.

Keywords: Adenoma; adrenal, Computed tomography, pheochromocytoma

I. Introduction

Pheochromocytomas are functionally active tumours derived from primitive rests of chromaffin cells. They are adrenal in origin in 90% of cases, although they can occur anywhere from base of skull to anus. 10% of these tumours are bilateral [1]. The incidence of bilaterality increases in paediatric patients and in patients with multiple endocrine neoplasia type 2 (Sipple Syndrome) [2,3]. About 10% of pheochromocytomas are malignant. These tumours undergo varied pathologic degeneration, hence presenting with a wide spectrum of imaging features. The epithet of 'kaleidoscope' explains the different appearances of tumour in different individuals. Often the large tumours are more cystic in nature in contrast to small solid ones. Their preoperative diagnosis is mandatory to avoid surgical emergency precipitated by hypertensive crisis due to increased catecholamine secretion. Patients may be completely asymptomatic, with up to 10% of cases being clinically silent [4]. These clinically silent pheochromocytomas are assumed to have little if any catecholamine secretion. Masses in such patients tend to be larger than hyperfunctioning tumors [5]. A 1979 study revealed that 50% of cases of pheochromocytoma were diagnosed at autopsy [6]. This study predates the burgeoning increase in the use of imaging during the past two decades, and many of these pheochromocytomas are now being encountered in vivo along with many other incidental adrenal tumors at CT [7].

II. Case Profile

A nine year old male child presented to the outpatient clinic with finding of a small mass felt in right flank. On clinical examination a small right sided renal mass was diagnosed. Laboratory investigations were within normal limits. Patient was further investigated with X-ray abdomen (Fig. 1), Ultrasonography and Computed tomography (Fig. 2). There was evidence of coarse calcification in right suprarenal region and lymphadenopathy on Ultrasonography suggesting the more common diagnosis of Adrenal tuberculosis. Contrast enhanced computed tomography of abdomen revealed a solid cystic lesion of right adrenal gland with enhancing solid part (in both arterial and portal venous phase) and foci of calcification (Fig. 3). Fifteen minutes delayed (post-contrast) scan revealed an absolute percentage washout (APW) of more than 60%. These findings corroborated with lipid-poor adenomas. Based on imaging diagnosis patient was taken up for surgery. On immunohistochemistry of excised specimen, the synaptophysin markers were identified consistent with final diagnosis of Pheochromocytoma.



Figure 1: Skiagram of abdomen showing coarse calcification in right lumbar region.



Figure 2: Plain axial CT abdomen showing coarse calcification within a soft tissue density in right adrenal gland



Figure 3: Contrast enhanced axial CT scan abdomen showing enhancing soft tissue mass (approximately 5x5 cm) with cystic areas and calcification in right adrenal gland.

III. Discussion

Pheochromocytomas are rare adrenal tumours found in only 10% cases in children. These tumours are often diagnosed clinically by findings suggestive of raised catecholamine levels in blood i.e. episodic hypertension and elevated urinary levels of catecholamine products. However, in clinically silent patients imaging diagnosis becomes important. Pheochromocytomas are usually large, heterogeneous masses with areas of necrosis and cystic change on Computed tomography which typically enhance avidly [8]. They may wash out similar to an adrenal adenoma, but they tend to have a greater enhancement in an arterial or portal venous contrast phase. They tend to enhance more on the portal venous phase than the arterial phase. 110 HU of enhancement on the arterial phase is compatible with pheochromocytoma; however, hypervascular metastases could be considered in an appropriate setting. Calcification is demonstrated in upto 7% of cases [9]. Most incidental adrenal lesions are detected at CT, with characterization of the lesions being predominantly dependent on attenuation parameters [1]. CT attenuation values have proved particularly valuable for characterizing adrenal adenoma, the most common adrenal tumor. Adenomas can often be differentiated from other masses owing to intracellular fat, which causes attenuation to decrease to less than 10 HU [10,11]. Most pheochromocytomas have an attenuation higher than 10 HU; rarely do they contain sufficient intracellular fat to have an attenuation of less than 10 HU [12].

Still, some pheochromocytomas could be incorrectly categorized as adenomas [11]. Pheochromocytomas, in typical inconsistent fashion, can demonstrate different and variable washout patterns and may again, therefore, be confused with either adenomas or metastases [12,13,14]. This inconsistent contrast material washout may be due to varied pathologic degeneration that produces abnormal capillary networks, which alter both enhancement and washout. It should be remembered, however, that there are some theoretical concerns about administering iodinated contrast material to a patient with pheochromocytoma. Despite reassuring reports of studies performed with low-osmolar nonionic contrast agents [15,16], current accepted practice would generally avoid the use of iodinated contrast material in a patient known to have a pheochromocytoma.

IV. Conclusion

Pheochromocytomas portray a perfect example of imaging kaleidoscope which should be investigated meticulously to avoid misdiagnosis and hence surgical complications. Complete work-up for pheochromocytoma should be taken up in cases of incidental suprarenal mass. Their unusual association with coarse calcification and lymphadenopathy confirms the rarity of the lesion and its variable presentations.

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