Adrenal Cyst-Diagnostic Dilemma: Case report

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Introduction

Adrenal cyst (AC) are rare lesions with heterogeneous origin. The most common type are endothelial cysts, followed by pseudocysts, epithelial and parasitic cysts. Malignant and functional adrenal tumors should be included in the differential diagnosis of AC as they can radiologically present as cystic lesions.

The size of the AC determines their clinical presentation. Small AC are clinically silent. Large AC presents with signs and symptoms either due to mass effect or intracystic haemorrhage.

Case report

A 57-year-old woman presented with episodes of right upper abdominal pain accompanied by headache, elevated blood pressure and vomiting. Abdominal ultrasound revealed right adrenal heterogenous mass 7 cm in size, with mass effect on adjacent organs, confirmed by CT scan as adrenal mass with internal septations and density >30HU, suspected for pseudocyst. Biochemical evaluation for functional adrenal mass was unremarkable. Notably, 24-hour urine metanephrines and vanilmandelic acid were within normal range on several occasions. Iodine-123 MIBG scintigraphy was not available at the moment of investigations.

Clinical suspicion for silent cystic pheochromocytoma (PCC) was established and the preoperative treatment with α and beta blockers was provided. Laparoscopic adrenalectomy attempt failed due to intraoperative hypertension (300/150 mmHg).

This event reinforced our concern for cystic PCC. An open adrenalectomy was performed three months later with no intraoperative hemodynamic instability.

Histopathologic analysis confirmed hemorrhagic epithelial cyst. The cyst wall contained epithelial cells on imunohistohemistry positive for AE1/AE3 and negative for podoplanin. Additionally the cyst wall included rim of fibrous and adrenal tissue (confirmed by chomogranin, sinaptofizin immunohistochemistry), lymphocytes and network of blood vessels. Within the cyst cavity presence of clusters of multinuclear giant macrophages (CD68+), hemosiderin and haemolyzed erythrocytes were present.

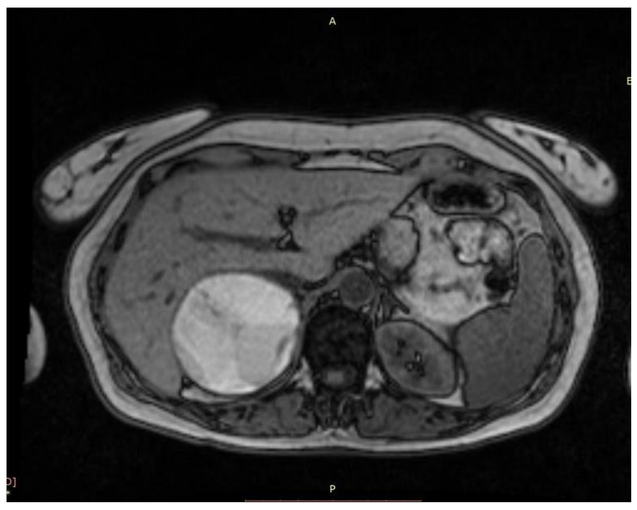

Figure 1. MRI image shows adrenal cyst with **v**ariable signal intensity on T1 weighted images High signal on T1 suggestive of the presence of blood

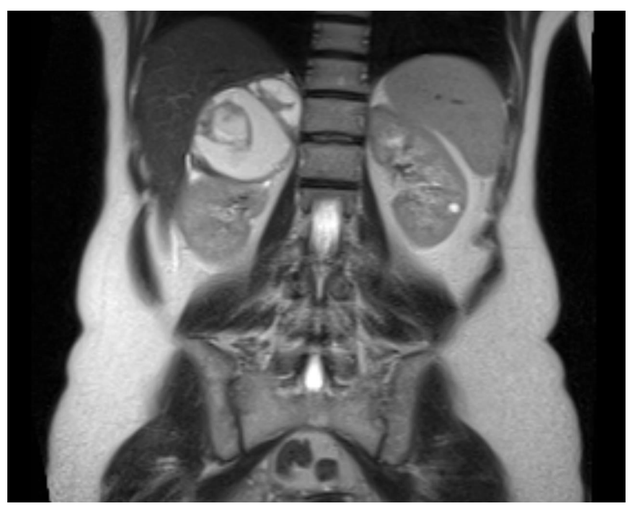

Figure 1. MRI image shows adrenal cystic lesion with high signal intensity on T2-weighted images, fluid-fluid levels and septation of the lesion..

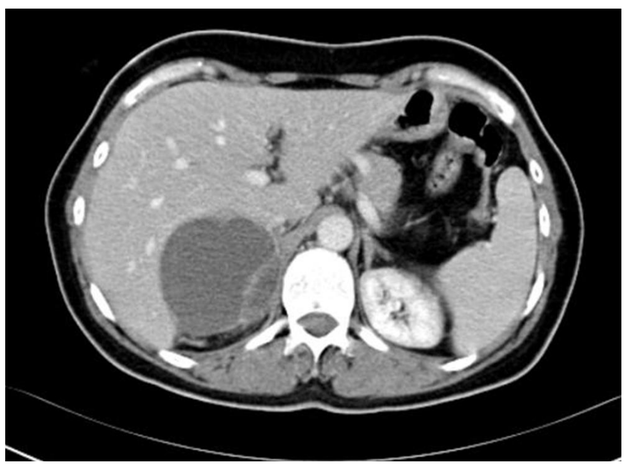

Figure 3. CT image with contrast shows contrast enhancement in the capsule and septation of adrenal cyst.

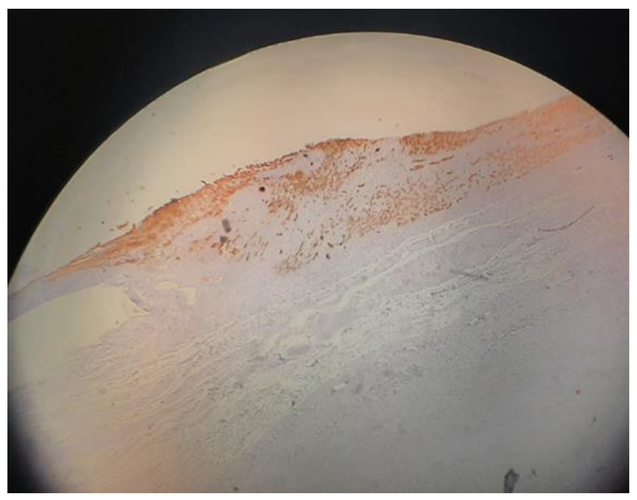

Figure 4. Histopathology analysis show cyst wall with epithelial lining positive for AE1/AE3 and granulation tissue within the cyst wall

Discusion

Functional adrenal tumors (i.e PCC), can radiologically appear as AC (pseudocyst) due to haemorrhage and cystic degeneration of the tumor. Additionally, silent cystic PCC lacks clinical presentation and biochemical diagnosis until surgical excision. The pathogenesis of cystic PCC is contributed to tumour outgrowing its vascular supply. Subsequent haemorrhage within the mass reduce the tumour cell population and entrapt catecholamine products inside the cyst.

Our assumption is that intraoperative hemodynamic instability in our patient could be as a result of coexistence of cystic PCC that initially presented during surgical manipulation.

We consider that, sufficient histopathological specimen was not provided due to intracystic haemorrhage and insufficient pathologic exploration of the large size cyst.

Another potential explanation for our patient intraoperative hemodynamic instability could be an adrenal haemorrhage and entrapment of cateholamines within the cyst that were ultimately released during surgical manipulation. The presence of blood vessels within the cyst wall and erythrocytes in the cyst cavity are in line with our assumption. Reported cases of hypertensive urgency in setting of adrenal haemorrhage support our viewpoint.

The literature has reported adrenal tumors in association with endothelial and pseudocyst but not with epithelial cyst, as it was shown in our case.

Concluison

A catecholamine storm and hemodynamic instability during surgery in patients with complex AC is possible scenario therefore preoperative treatment with alpha and beta blockers should be considered.

We consider long clinical follow up in our patient due to unresolved clinical presentation.

References

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