Case report

Schwannoma of the adrenal gland

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Abstract
Visceral schwannomas are extremely rare and are usually discovered incidentally on USG/CT-Scan. Primary schwannomas of the adrenal gland are extremely uncommon. It has been theorized that they originate from Schwann cells that insulate the nerve fibers innervating the adrenal medulla. Histopathological examination coupled with immunohistochemistry provides the definitive diagnosis. A 55 year old normotensive female presented with pain in the right loin since 5 months. Her renal parameters were normal. Contrast enhanced computed tomography of abdomen showed a well delineated 6.5 x 5cms mass at upper pole of her right kidney. 24-hour urinary metanephrine was slightly elevated—3.07mg/24hrs. A decline in Serum cortisol levels was observed following a dexamethasone suppression test (18.89nmol/l). Histopathological examination revealed a spindle cell tumor. Immunohistochemistry showed strong and diffuse positive staining for S-100 with negative expression for CD-117, desmin, CD-34, HMB-45, synaptophysin, chromogranin, cytokeratin, and SMA. Ki-67 index was 2%. A diagnosis of cellular schwannoma of adrenal gland was confirmed.

Key words: adrenal gland, immunohistochemistry, S-100, schwannoma

Schwannoma (Neurilemoma) is an encapsulated benign nerve sheath tumor usually originating from Schwann cells of peripheral, motor, sensory, sympathetic or cranial nerves. The tumors have a predilection for the head, neck, and flexor surfaces of the upper and lower extremities. Deeply situated tumors predominate in the posterior mediastinum and the retroperitoneum. Visceral schwannomas, especially of the adrenal gland, are extremely rare and are usually discovered serendipitously. Adrenal schwannomas are thought to arise from Schwann cells associated with the phrenic nerve, the vagus nerve, and the sympathetic trunk that innervate the adrenal medulla. A 55 year old female patient presented with pain in the right loin since 5 months. On evaluation she was normotensive, routine blood investigations, renal parameters were normal. 24-hour urinary metanephrine was slightly elevated—3.07mg/24hrs (1mg/24hrs). A decline in serum cortisol levels was observed following a dexamet}-
thasone suppression test—18.89nmol/l (171-536nmol/l). The abdominal ultrasound and CECT Abdomen showed well delineated 6.5 x 5cms homogenous mass at upper pole of Right kidney. A standard open adrenalectomy was performed without any complications. The postoperative period of the patient was uneventful.

Post operative gross pathological examination revealed a solitary adrenal tumour measuring 8 x 6 x 3cms with cut-section-grey tan to grey yellow to grey white.

On histopathological examination, the lesion showed compact spindle cell areas, densely populated, forming fascicular, and whorled growth pattern with eosinophilic cytoplasm and wavy, fusiform nuclei. Low mitotic activity was detected but atypical mitosis and necrosis were not observed. In contrast to classic form, this variant was devoid of Verocay bodies and Antoni B areas.

Immunohistochemical analysis revealed cells that were uniformly and strongly S-100 positive and were negative for CD117, desmin, CD34, HMB-45, synaptophysin, chromogranin, cytokeratin, and smooth muscle actin.

Discussion

Schwannoma is a benign, slow-growing, encapsulated neoplasm in which the principal component arises from neural crest cells and comprises differentiated Schwann cells in a poorly collagenized stroma. Schwannomas were first described by Verocay in 1908, with further sub-classification into two distinct histologic patterns performed by Antonini in 1920.

Adrenal schwannomas are typically found incidentally (‘adrenal incidentaloma’); however, patients may present with clinical symptoms secondary to the mass effect of the tumor. In review of literature by Mohiuddin Y et al, 13 out of 30 cases of adrenal incidentalomas presented with symptoms of abdominal or flank pain and discomfort. Tumors causing pain were greater than 5.0 cm in diameter and a single case with abnormal laboratory findings showed an elevation of urinary catecholamines.
that may be homogeneous. However, other cases in the literature have shown prominent cystic degeneration and calcifications. The majority of cases are firm, well-circumscribed rounded masses. They are described as tan yellow to grayish-white in appearance. Most are solid and homogeneous throughout.

A 2002 National Institutes of Health, science statement, on the management of adrenal incidentalomas concludes that lesions greater than 6 cm should be excised, those less than 4 cm with imaging characteristics that appear benign should generally not be resected, and those between 4 cm and 6 cm can be either closely observed or resected. However, non-secreting tumors with of size > 4cm can be excised with an added benefit of definitive diagnosis.

The differential diagnosis of adrenal incidentaloma can be extensive and ranges from cortical lesions such as adenoma and carcinoma to medullary lesions such as neuroblastoma, ganglioneuroma, pheochromocytoma, schwannoma and neurofibroma. Rare lesions like leiomyosarcoma and malignant peripheral nerve sheath tumors can also occur.

Adrenal schwannomas are thought to arise from the medulla, as there is continuity between the medulla and the tumor and an absence of a septum around the tumor. The masses often compress surrounding medullary and cortical tissue. Some adrenal schwannomas may grow very large and show ancient change, with hemorrhage, calcifications, and cystic change. This corresponds with the degenerative changes seen on imaging and grossly. Ancient change also may result in nuclear atypia. This nuclear atypia should not be confused with malignancy.

Schwannomas can be categorized as either conventional or cellular schwannomas based on their histologic features. Conventional schwannomas consist of alternating compact cellular areas and loosely textured paucicellular areas, termed Antoni A and Antoni B respectively. Antoni A area is composed of spindle shaped Schwann cells arranged in interlacing fascicles with nuclear palisading. In between two compact rows of well aligned nuclei, the cell processes form Eosinophilic Verocay bodies. Mitotic figures may be present (usually less than 5 per 10 high power field). Antoni B areas are far less orderly and less cellular. The spindle or oval cells are arranged haphazardly in the loosely textured matrix, which is punctuated by microcystic change, inflammatory cells, and delicate collagen fibers.

Cellular schwannomas were first described by Woodruff et al in 1981 and are highly cellular nerve sheath tumours arising mostly from spinal nerves. They consist entirely of Antoni A areas with intersecting fascicles of spindle cells and are devoid of hypocellular, Antoni B areas. They are also characterized by an absence of Verocay bodies and may have increased mitotic activity. They tend to develop more often in deep structures such as the posterior mediastinum and retroperitoneum. Immunohistochemistry of adrenal schwannomas shows strong and diffuse staining for S-100. They also display pericellular reactivity for collagen IV. They are typically negative for CD117, desmin,
CD34, HMB-45, synaptophysin, chromogranin, cytokeratin, and smooth muscle actin. This helps in ruling out the other differential diagnosis.

**Conclusion**

Adrenal schwannoma is a rare type of adrenal incidentaloma discovered usually on imaging or autopsy. Classifying this tumor can be challenging because imaging studies are nonspecific and many entities appear similar histologically. Because of its rarity, schwannoma occurring at this particular site can pose problems in diagnosis and should be considered as differential diagnosis and distinguished from other spindle cell lesions of the adrenal gland. Ancillary studies such as immunohistochemical analysis and electron microscopy help to provide a specific diagnosis.

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**References**


