CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 74 (2020) 173–176

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Adrenal schwannoma in a female patient; A rare benign neoplasm: Case report and literature review

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ARTICLE INFO

Article history:
Received 18 June 2020
Received in revised form 30 June 2020
Accepted 18 July 2020
Available online 26 July 2020

Keywords:
Schwannoma
Incidentaloma
Adrenalectomy
Rare
Benign
Tumor

ABSTRACT

INTRODUCTION: Schwannomas are rare, slow-growing, usually benign tumors that originate from myelin-producing Schwann cells. Adrenal schwannomas are an exceptionally rare subset of these tumors, with few cases reported in the literature.
PRESENTATION OF CASE: We present the case of a 44-year old female patient being evaluated for chronic abdominal pain at the outpatient clinic. Clinical and laboratory workup was unremarkable. An abdominal CT scan was performed, revealing a left suprarenal solid mass (5 × 6 cm). Surgical resection of the adrenal gland was performed, given the patient's symptoms, the size of the tumor, and its malignant potential. The patient completed the postoperative period satisfactorily, and her symptoms improved. Histopathological findings were compatible with a benign adrenal schwannoma.
DISCUSSION: Schwannomas generally appear in the head, neck and extremities, with the vestibulocochlear nerve being the most frequently involved site. Retroperitoneal schwannomas account for 1–5% of retroperitoneal masses and comprise only 1–3% of all schwannomas. Their incidence increases with age, from 4% in the general population, reaching 7% in patients over 70 years of age.
CONCLUSION: Adrenal incidentalomas represent a diagnostic challenge. Because of the malignant potential of large (> 4 cm) adrenal masses and the lack of characteristic findings using conventional imaging techniques and laboratory diagnostic tools, surgical excision with histopathology and immunohistochemistry analysis are required for definitive diagnosis and optimal management.

1. Introduction

Adrenal incidentalomas are adrenal masses that are discovered incidentally after performing radiologic examination of the abdominal cavity (CT, MRI) for evaluation of non-adrenal disease. Incidentalomas are relatively common radiologic findings, with a prevalence of up to 4% in the general population [2]. Schwannomas are rare, slow-growing, usually benign tumors that originate from myelin-producing Schwann cells. They usually arise in the skull, head, neck, extremities, and, much less frequently, in the retroperitoneal space [1–5]. We report the incidental finding and surgical management of an adrenal mass in a 44-year old female patient being evaluated for chronic abdominal pain.

*This work has been reported in line with the SCARE criteria [14].

2. Case report

A 44-year-old female patient came to the medical office with the chief complaint of chronic abdominal pain, with vague localization, mild intensity, and multiple failed therapeutic interventions. Medical history was unremarkable except for mild abdominal discomfort.

On physical examination, vital signs were within normal limits and no abnormalities were noted except for mild tenderness to palpation on the epigastric area.

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https://doi.org/10.1016/j.ijscr.2020.07.063

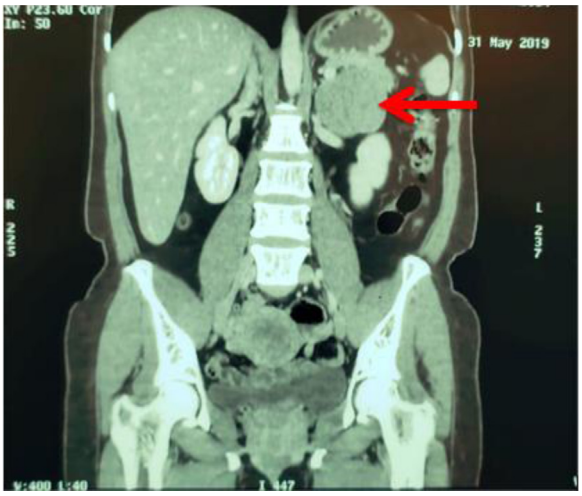

Fig. 1. CT scan image showing an Adrenal "Incidentaloma".

Abdominal CT scan revealed a solid, 5 × 6 cm lesion on the left adrenal gland and findings compatible with a hiatal hernia (Fig. 1).

Surgical resection of the adrenal mass was performed using a laparoscopic approach; a 5 cm mass was identified in the left adrenal gland. The tumor was carefully dissected with preservation of the rest of the gland, and the specimen was sent to pathology for analysis. Post-operative recovery was uneventful, and the patient was discharged 4 days after surgery.

Macroscopic evaluation of the mass showed a yellowish-white solid neoplastic lesion with a smooth homogeneous external capsule (Fig. 2).

Microscopic examination revealed neoplastic proliferation of fusiform cells arranged in interlocking bundles, a predominantly collagenous stroma with myxoid degeneration and inflammatory cells; the proliferating cells showed minimal pleomorphism, elongated nuclei with a regular contour, homogeneous chromatin and isolated inconspicuous nucleoli, without the presence of a significant number of mitosis figures (< 1 mitosis for every 10–40X fields). No areas of necrosis within the lesion were identified. Scarce adrenal tissue was identified in the periphery of the lesion (Fig. 3). Immunohistochemistry was performed, with diffuse and intense positivity for S-100, SOX-10 and CD34. Staining was negative for STAT-6 (fibrous tumor), CD117 (gastrointestinal stroma) and smooth muscle actin (leiomyoma). Ki67 proliferation index was 1%. These findings correspond to a benign nerve sheath tumor, or Schwannoma (Fig. 4).

3. Discussion

Schwannomas were first described by Verocay in 1908. In 1920, Antonini sub-classified them into 2 distinct histologic patterns: types A and B. Schwannomas are benign, slow-growing, encapsulated neoplasms [9–11]. Generally, these tumors appear in the head, neck, upper and lower extremities, with the vestibulocochlear nerve being the most frequently involved site. Retroperitoneal schwannomas account for 1–5% of retroperitoneal masses and comprise only 1–3% of all schwannomas. On gross examination, schwannomas are encapsulated tumors that vary from firm and homogeneous masses to fluctuant cysts [1–8]. Pathophysiology is not well understood; current theories propose that adrenal gland schwannomas originate from Schwann cells that insulate the nerve fibers (phrenic, CN X, sympathetic chain) innervating the adrenal medulla [2–5].

Most functional tumors of the adrenal gland can guide diagnosis by the symptoms they produce and can be further evaluated through laboratory exams, such as blood chemistry and urinary tests. Non-functional tumors do not share this characteristic, and are most likely to be diagnosed incidentally through CT [12]. Such is the case of adrenal schwannomas, which are commonly detected incidentally as a defined oval mass with cystic degeneration or calcification. These findings may also be suggestive of malignancy [9]. MRI can also help identify an adrenal schwannoma, but its findings are not particularly specific; the tumor may appear as a low intensity mass in T1 with heterogeneous high intensity in T2 [9].

The preoperative diagnosis of an adrenal schwannoma is almost impossible to achieve. Even when suspected after radiological examination, histopathological confirmation is required. It should also be taken into account that up to 50% of neurofibrosarcomas or malignant schwannomas are associated with neurofibromatosis type 1, making excisional biopsy an indispensable tool for the accurate characterisation of these tumors [1–5]. Because of the risks and limitations of fine needle aspiration biopsy, it should only be

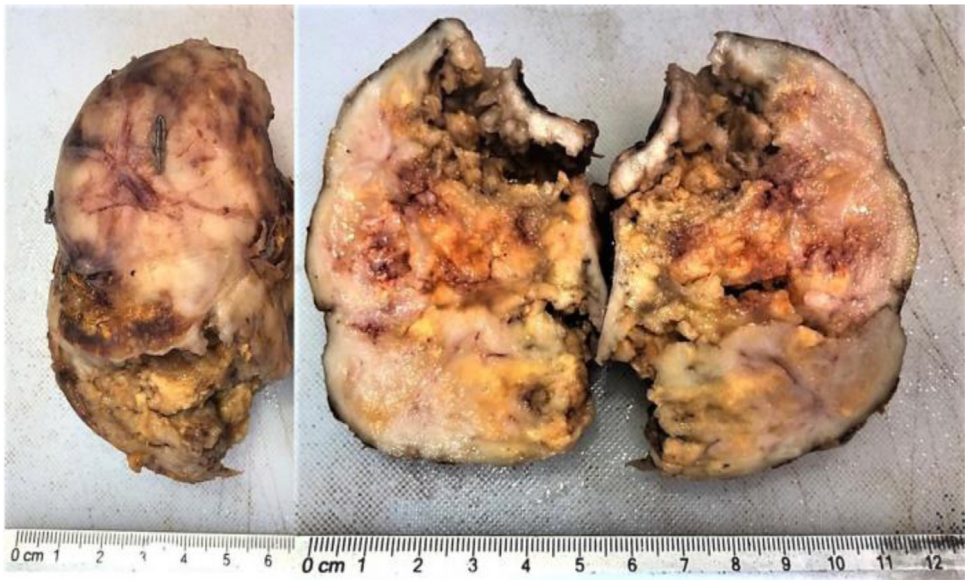

Fig. 2. Surgical specimen showing a yellowish-white solid neoplastic lesion with a smooth homogeneous external capsule.

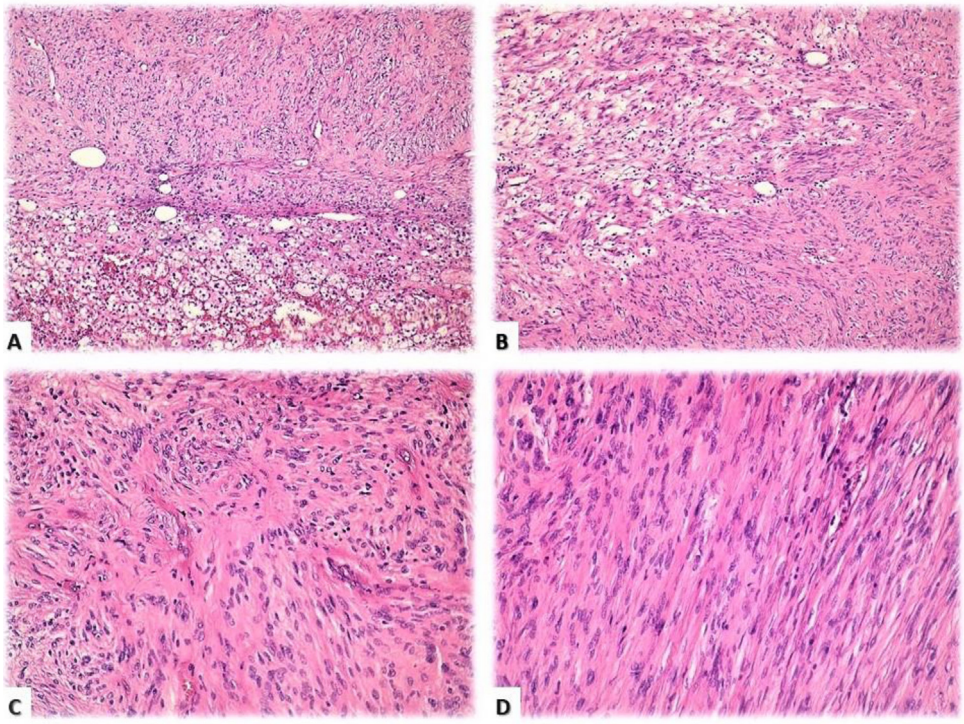

Fig. 3. H& E slides showing proliferation of fusiform cells arranged in interlocking bundles, a predominantly collagenous stroma with myxoid degeneration and inflammatory cells; the proliferating cells showed minimal pleomorphism, elongated nuclei with a regular contour, homogeneous chromatin and isolated inconspicuous nucleoli, without the presence of a significant number of mitosis figures (< 1 mitosis for every 10–40X fields). No areas of necrosis within the lesion were identified. Scarce adrenal tissue was identified in the periphery of the lesion.

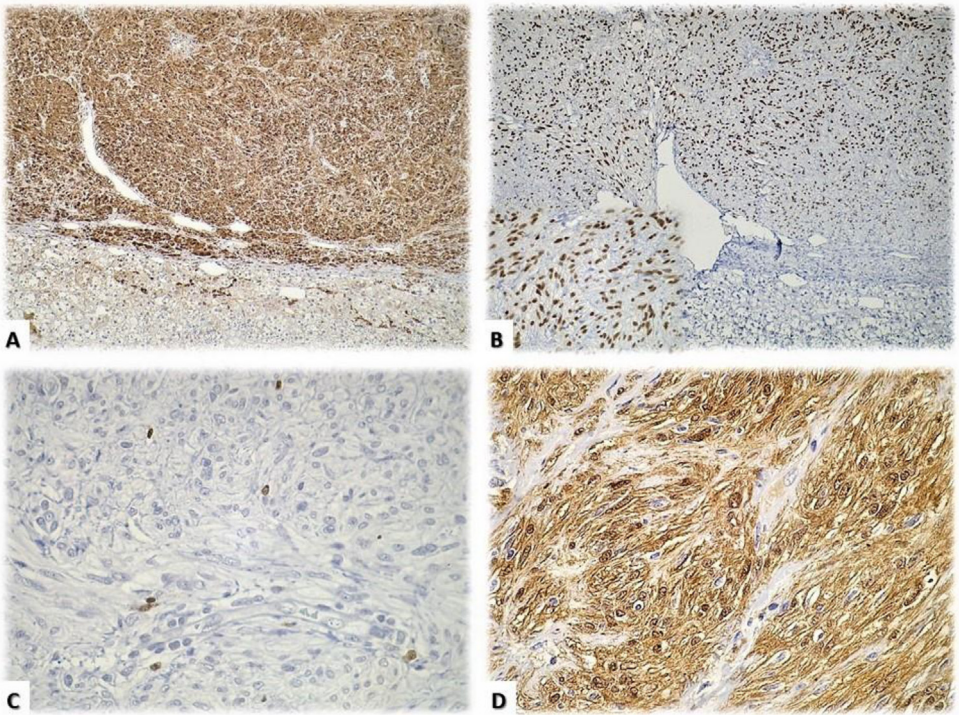

Fig. 4. Immunochemistry showing diffuse and intense positivity for S-100, SOX-10 and CD34.Staining was negative for STAT-6 (fibrous tumor), CD117 (gastrointestinal stroma) and smooth muscle actin (leiomyoma). Ki67 proliferation index was 1%.

considered in patients with metastasis or a previous diagnosis of carcinoma [2].

Recovery after surgery is usually achieved without complications. The possibility of malignancy or recurrence must always be considered even with a previous diagnosis of benign schwannoma [22]. The current recommendation for adrenal non-functional masses is surgical removal if $\geq$ 4 cm in diameter. Complete excision must be performed and sent for definitive diagnosis to

pathology with histological and immunohistochemical examination.

Inmunochemistry analysis is always performed for diagnosis; Schwannmas stain positively for S-100 antigen, vimentin, collagen IV and laminin and negatively for keratin, desmin, actin, CD34 and CD117 [13].

4. Conclusion

Adrenal schwannomas are an exceptionally rare differential diagnosis of the incidentally discovered adrenal mass, representing a diagnostic challenge. Because of the malignant potential of large (> 4 cm) adrenal masses and the lack of characteristic findings using conventional imaging techniques and laboratory diagnostic tools, surgical excision with histopathology and immunohistochemistry analysis are required for definitive diagnosis and optimal management.

Funding

There was no sponsorship for this study.

Ethical approval

There was no need for ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

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Felix Alejandro Perez Trsitan MD: Data collection, data analysis, writing the paper.
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Guarantor

Jesus Flores Armenta MD.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of Competing Interest

There was no conflict of interest.

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