Rare extra-adrenal paraganglioma mimicking a painful Schwannoma

Case report

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INTRODUCTION:
Paragangliomas originate from chromaffin tissue primarily found in the Zuckerkandl body. A 53 years old man, was investigated with abdominal MRI for chronic backache, that had lasted for 2 years.

CASE REPORT:
MRI revealed an incidental mass in para-aortic region, at the adrenal lodge, with characteristics of a solid nodular mass, which did not seem to arise from the adrenal gland. An abdominal CT showed a mass localized prior to left adrenal gland; the radiologist reported this mass as a malignant peripheral nerve sheath tumor (Schwannoma).

During his stay in hospital, tumoral markers chromogranin and neuron-specific enolase were dosed, with a concentration of 187.00 ng/mL and 7.7 ng/mL. Patient’s back pain was treated first with ketorolac, without any resolution of the problem, then with ketorolac plus tramadol in elastomeric pump, again with no relief, and finally with a continuous infusion of morphine. Since the exact nature of the neoplasm was not known and a CT guided biopsy was not technically feasible to do, a laparoscopic excision of the mass, with transperitoneal anterior submesocolic approach, as well as for the left adrenalectomy, was planned. After surgery, the patient no longer needed morphine. The postoperative course was uneventful.

RESULTS:
Histopathological findings consisted of periadrenal paraganglioma. Extra-adrenal paragangliomas are rarely recognized during preoperative study. This is often due to its capacity of mimicking other type of tumors.

CONCLUSIONS:
Paragangliomas could be successfully resected by laparoscopy, they are difficult to distinguish from other tumor and they should be investigated even if preoperative findings argue for something else.

KEY WORDS: Laparoscopic adrenalectomy, Paraganglioma, Schwannoma

Introduction

The incidence of paragangliomas is 1/100.000, and are approximately ten percent of catecholamine secreting tumors. Paragangliomas originate from chromaffin tissue primarily found in the Zuckerkandl body, the kidneys, the sympathetic plexus of the urinary bladder, and the heart or in the sympathetic ganglia of the head or neck.

Due to the possibility of catecholamine's secretion, patients with paragangliomas may present with signs and symptoms including hypertension, palpitations, tremor, pallor, headache and tachyarrhythmia. Catecholamine-secreting paragangliomas are only 25%. The remainder present with an abdominal mass only and no evidence of hypertension.

Case report

A 53 years old man, followed up for a previously treated Hodgkin’s lymphoma in 1993 and with hypercholesterolemia/hypertriglyceridemia, was investigated with...
abdominal MRI for chronic backache that had lasted for two years. The patient tried to relieve the back pain (VAS Score 8), unsuccessfully, with several types of non-steroidal anti-inflammatory drugs (NSAID). MRI revealed an incidental para-aortic solid nodular mass (29x25mm) located within the left adrenal lodge, which did not seem to arise from the left adrenal gland. CT scan (Fig. 2) showed the mass to be localized anteriorly to the left adrenal gland and was reported as a malignant peripheral nerve sheath tumor (Schwannoma) or, alternatively, as a partially colliquated lymph node. Biopsy, due to its difficult location, was not performed. The patient did not report headaches, excessive sweating, palpitations, hypertension, or any other systemic symptom. During his stay in hospital, tumor markers chromogranin and neuron-specific enolase (NSE) were 187.00 ng/mL (n.v. <90.00 ng/mL) and 7.7 ng/mL (n.v. <12 ng/mL), respectively.

The patient's back pain was treated with ketorolac first, with no relief, then with ketorolac plus tramadol in elastomeric pump, again with no relief, and finally with a continuous infusion of morphine, which was successfully.

**Treatment**

Since the exact nature of the neoplasm was not known and a CT guided biopsy was not technically feasible, a laparoscopic excision of the mass was planned, with a transperitoneal anterior submesocolic approach, as commonly performed by the authors for left adrenalectomy.

Surgery was performed under general anesthesia. A nasogastric tube and a urinary catheter were positioned. The patients was monitored with an intra-arterial radial artery catheter for blood pressure measurement and a central venous catheter (internal jugular access) to monitor changes in central venous pressure and for rapid introduction of liquids, if necessary. Pneumoperitoneum was established with a Veress needle at the umbilicus. Pneumoperitoneum at 13 mmHg pressure was obtained with carbon dioxide insufflator adjusted at 30 l/minute. Four 12mm trocars (Fig. 1) and a 45° optic were used. This approach was performed with the patient supine, in anti-Trendelenburg position and with the operating table rotated 30° towards the opposite side of the lesion, to facilitate exposure of the surgical field. The surgeon stands on the side ipsilateral to the lesion.

The first trocar was inserted to the left of the midline above the umbilicus (Fig. 1). A second trocar was inserted on the right midclavicular line below the right costal arch. A third and fourth trocar were placed one on the left midclavicular line above the transverse umbilical line and one on the left anterior axillary line, respectively.

The transverse mesocolon was raised by the assistant withatraumatic forceps introduced through the right midclavicular trocar to expose the first jejunal loop at the ligament of Treitz and to identify the inferior mesenteric vein. The posterior peritoneum was opened at the insertion of the transverse mesocolon on the lower edge of
the pancreas, between the first jejunal loop and the inferior mesenteric vein. After Toldt’s fascia identification, the dissection proceeded posteriorly along the retro-pancreatic plane, elevating the body of the pancreas. Then Gerota’s fascia was opened to identify the superior margin of the left renal vein. This was prepared latero-medi ally until its junction with the inferior adrenal vein, which was adherent the mass. The left adrenal vein was prepared and divided between clips (AcuClip, Tyco / Healthcare, Norwalk, Connecticut, USA) to allow access to the mass. After adrenal gland division, the medial subdiaphragmatic connections of the mass with the surrounding anatomical structures were divided with a radiofrequency vessel sealing device (LigaSure™ tissue fusion, Covidien, Mansfield, Massachusetts, USA).

Monopolar electrocautery was selectively used for hemostasis. Once released, the mass was removed inside an extraction bag. The residual space was filled with hemostatic material (Floseal, Baxter Healthcare Corporation, Deerfield, Illinois, USA) and a drainage was left in place. After surgery, the patient no longer needed morphine and the pain was easily controlled with paracetamol, suspended on POD 3. The postoperative course was uneventful.

The resected specimen consisted of a brownish tissue of 5x3x2 cm, with an area of central colliquation. Histological examination revealed a polygonal cells’ nests mass, which presented a round- to oval-cell predominant smear with occasional striking anisonucleosis, free of histological anomalies. Mitosis or necrotic areas were not seen. Immunohistochemical stainings have reported diffuse positivity with chromogranin, synaptophysin and S100 protein; CKAE1-AE3 and melan-A were negative. The proliferation index was found to be 2-3%. Histopathological findings were consistent with the diagnosis of periadrenal paraganglioma.

Follow-up

The patient is alive, free of symptoms and has been followed up for 15 months. Follow up abdominal CT and MRI scans were repeated, both of them were unremarkable.

Discussion/Conclusion

Extra-adrenal paragangliomas are rarely recognized preoperatively. This is often due to its capacity of mimicking other types of tumors. 6-15 Magnetic resonance imaging (MRI) and computerized tomography (CT) are highly sensitive in detection of small tumors but are not specific for paragangliomas. 16-17 MIBG scintigraphy is specific for paraganglioma but is less sensitive than MRI and CT. 18 Due to normal plasma and urine catecholamine levels and to CT and MRI findings that mimicked a malignant peripheral nerve sheath tumor, a MIBG scintigraphy, however, was not performed.

Paragangliomas are difficult to distinguish from other tumors. Any extra-adrenal mass should be investigated even if the preoperative findings argue for something else. In this patient, the paraganglioma mimicked an adrenal mass and it was treated with the same laparoscopic approach that is commonly employed by the authors to treat adrenal masses, with good results. The submesocolic approach was found to be safe and feasible to treat this peri-adrenal mass.
Gli Autori credono che i paragangliomi possano essere resecati con successo tramite approccio laparoscopico, che essi sono difficili da distinguere altri tipi di tumore e che debbano essere sospettati anche se i risultati preoperatori depongono per qualcos’altro.

References