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Scientific Letter

Gastric Hybrid Schwannoma/Perineurioma☆

Schwannoma híbrido gástrico/perineurioma

Benign peripheral nerve sheath tumors include neurofibromas, schwannomas and the less common perineuriomas. New cases showing features of more than one histologic type have been recognized in recent years, with the great majority arising in peripheral nerves of extremity and trunk. Pathogenesis is poorly understood and immunohistochemistry is essential to confirm the diagnosis. Hybrid tumors involving the gastrointestinal tract are extremely rare. We present a case of schwannoma/perineurioma of the stomach.

A 76-years-old Caucasian woman was referred to our Department with a 2 months history of abdominal pain, appetite and weight loss. Medical history included appendectomy, thyroidectomy for multinodular goiter, and atrial fibrillation. Physical examination was unremarkable and no sign of neurofibromatosis was identified. Hemoglobin level was 8.5 g/dl and common tumors markers were within the normal range. Esophagogastroduodenoscopy was negative. Multi-detector Computed Tomography scan showed a 7×6.5 cm isodense exophytic mass arising from the greater curvature of the stomach and abutting on the pancreatic tail and splenic hilum. At Magnetic Resonance Imaging it appeared irregular, mainly hipointense both in T1 and T2, with homogeneous enhancement after contrast administration (Fig. 1). No sign of hemorrhage, cystic degeneration or necrosis was detected. A presumed diagnosis of gastric mesenchimal tumor was made. Patient underwent wedge resection of the stomach with distal splenopancreatectomy. In the postoperative period a left subphrenic abscess occurred, requiring CT-guide percutaneous drainage and antibiotic treatment and the patient was discharged 12 days later. Macroscopically the tumor was a 7 cm×4 cm×4 cm nodular lesion, grossly well circumscribed lesion, with firm consistency and whitish cut surface. Microscopically the tumor showed a storiform growth and collagenous stroma dominant architectural features typical of perineurioma but many closely intermingled large spindle cells with plump tapered nuclei and eosinophilic cytoplasm typical of schwannoma were also observed (Fig. 2a). No significant nuclear atypia, mitotic activity or necrosis in either component was observed. Immunohistochemical analysis revealed the fascicular component diffusely positive for S100 (Fig. 2b), while the proliferation of cells was positive for Epithelial Membrane Antigen (EMA) (Fig. 2c) and Glut-1 (Fig. 2d). Others markers including CD34, smooth muscle actin, desmin, DOG-1 and CD-117 were negative, in agreement with the diagnosis of gastric hybrid schwannoma/perineurioma. The patient is currently alive without evidence of recurrence 3 years after surgery.

Benign peripheral nerve sheath tumors include a spectrum of lesions with well-defined clinicopathological features allowing a clear diagnosis. Those involving the gastrointestinal tract are rare, representing only ≤5% of all mesenchymal tumors and differential diagnosis with gastrointestinal stromal tumors GIST or smooth muscle tumors may be difficult without surgical excision.[1] Due to advances in molecular biology, a subset of tumors demonstrating morphologic features overlapping between different entities have been increasingly described and included in the 4th edition of the

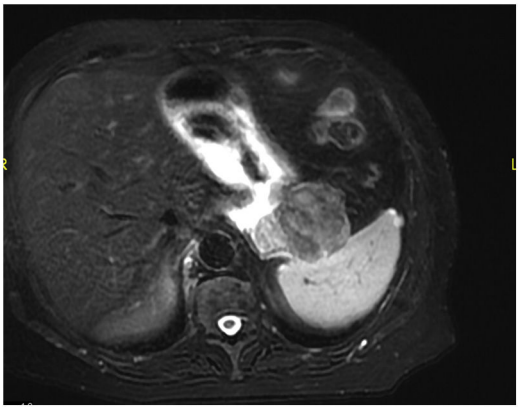

Figure 1 – Magnetic resonance imaging showing homogeneous enhancement of the lesion, appearing mainly hypointense both in T1 and T2.

☆ Please cite this article as: Carboni F, Valle M, Covello R. Schwannoma híbrido gástrico/perineurioma. Cir Esp. 2019;97:292–294.

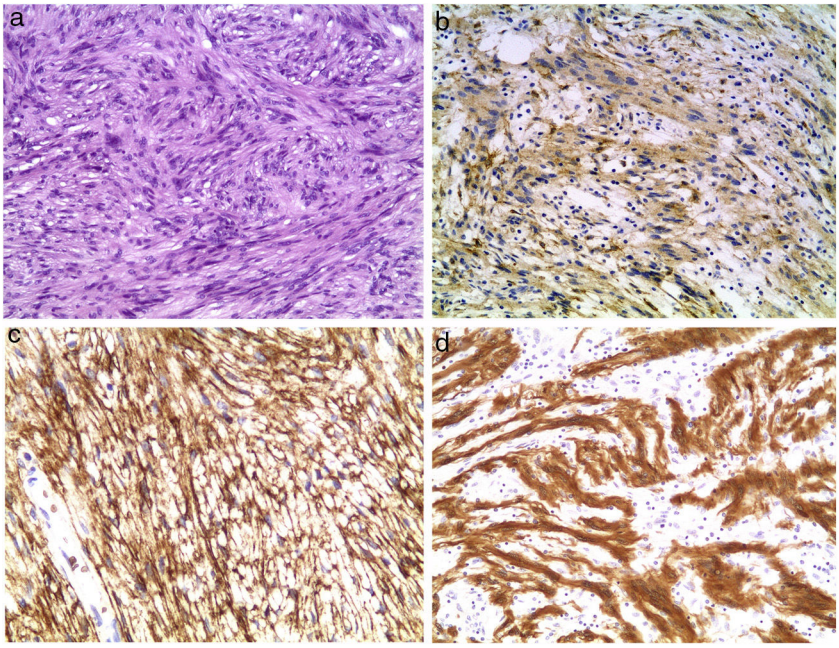

Figure 2 – Histological sections showing features typical of both perineurioma and schwannoma (H&E ×100) (a). Immunohistochemical analysis positive for EMA (×100) (b). Immunohistochemical analysis positive for Glut-1 (×200) (c). Immunohistochemical analysis positive for S100 (×200) (d).

World Health Organization Classification of Tumors of Soft Tissue and Bones.[2] They exhibit an abrupt transition and/or intimate admixture of cells type and hey have been termed ''hybrid'' peripheral nerve sheath tumors.[2–5] The pathogenesis of these lesions is poorly understood, but it might be associated with localized microenvironmental change or clonal genetic alteration in primitive tumor cells.[3] Tumors occurring in the gastrointestinal tract can be extremely difficult to classify and diagnostically very challenging. Immunohistochemical analysis is essential to confirm the dual differentiation of these tumors and it has been suggested that the routine use of immunohistochemistry may increase their frequency.[6] The co-existence of benign peripheral nerve sheath tumor with distinct schwannoma and perineurioma features has been reported but extraneural occurrence is extremely rare. Schwannomas typically show diffuse, strong expression of S100 protein. Epithelial membrane antigen is the most widely used marker and stains for most perineuriomas, with additional markers like Claudin-1 and Glut-1 being increasingly used.[5,7–9] Neither of these markers is entirely specific for perineural differentiation and they are best used as a part of a multi-antibody panel.[1,5,10] Differential diagnosis includes gastrointestinal stromal tumors, other benign peripheral nerve sheath tumors and low-grade malignant peripheral nerve sheath tumors.[2–5] The diagnosis of hybrid peripheral nerve sheath tumor is made in the presence of two distinct histological areas and is confirmed by different patterns of HIC staining of the two components. To the best of our knowledge, only another case has been documented in the stomach.[8]

Concerning treatment, complete surgical excision is curative since the behavior of such tumors is usually benign, but one recurrence after incomplete resection and two cases of malignant transformation have been reported.[11]

Conflict of Interest

None.

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2173-5077/

Melanoma of the Intestinal Mucosa: A Rare Cause of Obstruction☆

Melanoma mucoso intestinal: una rara causa de obstrucción

A 62-year-old woman, who had a history of uterine fibroids requiring hysterectomy and double adnexectomy at age 44, came to the emergency department of our hospital due to a 3-day history of emetic syndrome associated with altered bowel habits over the previous 2 months and weight loss of 12 kg.

On examination, the patient presented a slightly distended, soft abdomen with discomfort in the right iliac fossa but no peritonism. Bowel sounds were increased, and the digital rectal examination was normal.

Lab work showed hemoglobin levels of 9.2 g/dL, 15,000 leukocytes and a prothrombin activity of 73%; the remaining parameters were normal. Abdominal radiography demonstrated distended intestinal loops and the absence of gas in the colon and rectum. Abdominal CT scan identified 2 intestinal masses that were causing intestinal intussusception and mechanical obstruction (Figs. 1 and 2).

Urgent surgery was indicated, and exploratory laparoscopy revealed 2 intestinal loops with stenosis caused by tumors and retrograde dilation. No free fluid or space-occupying lesion in the liver were observed. Using an assistance incision, the intestines were eviscerated, an 80-cm section of the jejunum-ileum was resected and a side-to-side reinforced anastomosis was created mechanically.

The postoperative period was uneventful, and the patient was discharged on the sixth day.

The pathology study reported the presence of 2 lesions, both compatible with malignant melanoma of the synchronous intestinal type, with ulceration present. These lesions affected the mucosa and muscular layers. The proliferative index was high; maximum Ki-67 was 63%. Lymphovascular invasion was detected. The immunophenotype of the cells was vimentin, HMB-45 and INI1, positive diffuse; S-100 and SOX-10, positive focal; CKAE1/AE3, negative. No involvement was identified in the 12 lymph nodes studied.

The patient was evaluated by the Dermatology Unit, which ruled out malignant melanoma of cutaneous origin.

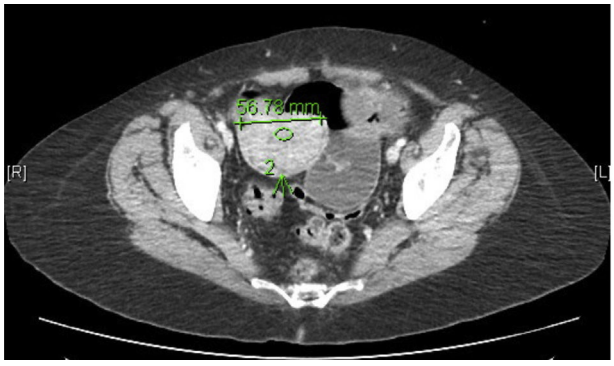

Fig. 1 – Intestinal mass and secondary intussusception.

☆ Please cite this article as: García-Romera Á, Morales Hernández A, Medina-Arana V, Bravo Gutiérrez A, Alarcó A. Melanoma mucoso intestinal: una rara causa de obstrucción. Cir Esp. 2019;97:294–295.