Not ery small bowel submucosal mass is a GIST
A rare case of small bowel schwannoma

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INTRODUCTION: Schwannomas are slow-growing tumors, rarely occurring in the GI tract. When found, their location is mostly in the stomach. Presentation in the small bowel is extremely rare, anecdotal.

CASE REPORT: We present the case of a 47-year old male who underwent laparoscopic elective left hemicolectomy for recurrent diverticulitis.

RESULTS: At surgery an exophytic mass originating from the first jejunal loop was identified and resected, under the suspicion of a GIST. Post-operative pathologic report was consistent with the diagnosis of jejunal schwannoma. Unfortunately resection margins were positive and the patient was taken back to the OR for an additional jejunal segmental resection.

CONCLUSIONS: GI schwannomas have excellent prognosis after surgical resection. It is important to differentiate them from GISTs, which may have a malignant behavior in 10-30% of the cases. To ensure complete surgical resection with negative margins is mandatory and is the curative treatment of choice.

KEY WORDS: GIST, Jejunum, Submucosa Minimally-invasive surgery, Schwannoma, Small bowel
unremarkable. In particular he had no history of epigastric pain or discomfort nor dysphagia to solids or weight loss. He had already visited the ER department of another hospital a few weeks before with the typical symptoms of diverticulitis and had been treated with antibiotics. Abdominal CT scans and colonoscopy confirmed severe sigmoid diverticulitis.

Laparoscopic hemicolecction was offered to the patient. At surgery, after completion of left colon resection, when we were closing the mesocolic defect, close to the angle of Treitz, with titanium clips as usual, we identified an exophytic mass originating from the first jejunal loop. We used an endovascular gastrointestinal anastomosis (GIA) stapler across the base of the mass (1x1 cm), ensuring there was no residual stricture in the jejunal lumen (Fig. 1). The specimen was retrieved through an endo-catch bag, via the umbilical port and sent to the pathologist for analysis. We suspected a GIST. The rest of the abdominal cavity, already explored at the beginning of the operation, was again visualized and there was no evidence of abnormalities. The recovery was uneventful and the patient was discharged home on post-op day 7. Histological examination revealed that the resected mass consisted of spindle cells with focal cellular atypia and lymphocytic cuffing at the periphery of the tumor (Fig. 2). The neoplastic cells were immunoreactive for S-100 protein (Fig. 3), but lacked immunoreactivity for, CK8-18, CD-117, CD-34, smooth muscle actin (SMA), HMB45, MART1, CK8-18 (Cam 5.2). The proliferation index (Mib-1) was 5-10%. The histopathologic features and immunohistochemical stainings were consistent with the diagnosis of jejunal schwannoma. Unfortunately spindle cells were documented at the resection margins. After discussion with the multidisciplinary oncological team and with the patient, we decided to take the patient back to the OR and let him have a laparoscopic jejunal segmental resection with termino-terminal anastomosis within two months from the first operation. The patient had a second uneventful recovery.

Discussion

Schwannomas (neurinomas or neurilemmomas) are benign tumors originating from Schwann cells. The may present anywhere along the peripheral course of nerves, but usually are rarely located in the GI tract and outnumbered by other submucosal mesenchymal tumors, such as GISTs (50-100:1) 9. Predominantly they occur in patients in their 60s, usually in the stomach and esophagus or occasionally in the intestines. They present with a spherical or ovoidal shape and may vary in diameter, on average 3 cm 34. Usually, they are covered by normally-appearing intact mucosa and grossly can resemble GISTs. On immunohistochemistry they show positivity for vimentin, S-100 protein and negativity for CD-34, CD-117, α-smooth muscle actin and desmin. These are the key elements to make the post-operative,
histopathological differentiation with GISTs, from which they are virtually indistinguishable when detected at CT scans or when found incidentally intraoperatively. Even when located in the stomach and identified preoperatively, biopsies are usually negative, given their submucosal location and the diagnosis is based on the anatomo-pathological and immunohistochemical analysis performed on the removed specimen. No malignant counterpart of this lesion has been described so far. However, it is important that resection be complete with no resection margins involved by the schwannoma. Recurrent disease has been described only following incomplete resection. Given the slow growth of schwannomas, their presentation is usually incidental, with aspecific complaints by the patient, including dyspepsia and epigastric pain when located in the stomach and constipation or rectorrhagia when the location is in the colon. Rectal location presenting with bleeding and being misdiagnosed with rectal cancer has also been described. Presentation of schwannoma in the jejunal loops may vary from asymptomatic to severely symptomatic with repeated episodes of bleeding requiring arteriography of the mesenteric artery for a precise localization of the bleeding. In our case, the patient had been studied with abdominal CT scan and colonoscopy for recurrent diverticulitis and no suspicion of submucosal lesion had been raised on pre-operative evaluation, nor he had any other aspecific symptom prompting further radiological or endoscopic examination. In fact the patient didn’t even have GI bleeding, which could be related to the submucosal growth compromising mucosal blood supply of the jejunum. When we completed the left hemicolectomy and were reapproximating the mesocolic defect, we noticed a jejunal ‘bulge’ and clearly suspected a GIST. Given the location of the lesion in one of the first jejunal loop, we decided for a wedge tangential resection, to allow for complete excision of the mass, preservation of the lumen caliber and prevention of anastomotic leak following segmental resection for a mass of unknown nature. The post-operative report prompted a multidisciplinary discussion with oncologists and pathologists. We revised the medical literature and clearly was evident that our case was anecdotal, more than just rare. When pre-operative examination or intraoperative findings suggest the possible presence of a GIST, a schwannoma, similarly located in the submucosa, should be suspected and clear margins of resections should be always obtained. However, we have to admit that, given the paucity of cases of GI schwannomas, the index of suspicion in the medical community is very low and could not be criticized. A preoperative radiologic differentiation between GIST and schwannoma is not easy: typical, distinctive, unique radiological features are lacking, and data from the medical (radiological or endoscopic literature) are scarce. Of the most important criteria to differentiate a schwannoma from a GIST is the density of the mass; internal structure is solid and homogeneous in the first case and it presents a central area with hemorrhage or necrosis and a peripheral enhancement in the second. Often different radiological exams, including MRI, are required to add information although inconclusive. However, when identified preoperatively, it is not easy to distinguish certainly GIST from schwannoma and often it is necessary to wait the results of the pathological examination. A post-operative differentiation of schwannoma form GIST is necessary: the latter bares a malignant behavior in up to 30% of the cases. To ensure complete surgical resection with negative margins is mandatory, is the curative treatment of choice, irrespective of schwannomas’ location and provides an excellent prognosis for these benign tumors.

Riassunto

Gli schwannomi sono dei tumori solidi a lenta crescita, che raramente si localizzano nel tratto gastro-intestinale. Quando vengono identificati, solitamente la loro localizzazione è gastrica. Il riscontro di queste lesioni a livello del piccolo intestino è estremamente raro, se non anecdotico.

In questo articolo, presentiamo il case-report di un paziente di 47 anni che era stato operato di emicolectomia sinistra per diverticoliti ricorrenti. In corso di intervento, veniva individuata una massa esofatica ad origine dalla prima anse ileale. Tale lesione veniva asportata nel sospetto si trattasse di un GIST. L’esame istologico del pezzo operatorio ha aveva dimostrato trattarsi di schwannoma digiunale. Sfortunatamente i margini di resezione erano positivi per cui si è reso necessario ripetere il paziente in sala operatoria per una resezione digiunale addizionale.

Gli schwannoma del tratto gastro-intestinale hanno un’ottima prognosi dopo resezione chirurgica. È importante differenziarli dai GIST, che possono avere comportamento maligno nel 10-30% dei casi. Una resezione chirurgica completa con margini negativi è necessaria e rappresenta il trattamento curativo di scelta.

References


