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Laparoscopic Resection of Ancient Schwannoma of the Small Bowel

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Abstract: ***Schwannomas are benign neurogenic tumors that arise from Schwann cells that line the sheaths of peripheral nerves. Schwannomas are commonly located in the soft tissues of the head and neck, extremities, mediastinum, retroperitoneum, and pelvis, but they are very rare in the mesentery. We report on the case of an ancient schwannoma of the small bowel arising in a 27-year old asymptomatic woman, which was resected successfully using laparoscopic surgical technique. The aim of this report is to recognize the possibility of schwannomas in the differential diagnosis of abdominal slowly growing tumors.***

1. Introduction

Schwannomas (or neurilemmomas) are solitary and generally benign tumors originating from the cells of the peripheral nerves sheath. The majority of the cases reported in the literature occur in the head and neck region. The "*ancient*" variant of schwannomas is an even more relative rare subtype. Histopathological features tend to classify this variant as a result of reactive/degenerating processes occurring to an inveterate lesion. We report on the case of an ancient schwannoma of the small bowel arising in a 27-year old asymptomatic woman, paying particular attention to the differential diagnosis with other benign or malignant tumors and increasing the consciousness that these lesions may also develop in other less common regions.

2. Case Report

27 years-old female presented with lower abdominal mass accidentally discovered. The patient was absolutely asymptomatic. Previous TC scan localized the lesion in the ileal mesentery. Previous ultrasonography was not helpful, showing non specific features. For this reason, the patient was subjected to a magnetic resonance to better characterized the lesion. Contrast enhancement showed notable vascularization, so that the result of the magnetic resonance tended to classify the lesion as (mesenterial desmoid or GIST). In order to shed light on the nature of the lesion, a decision was made to undertake the patient laparoscopic en-block resection of the lesion and the adjacent ileal mesentery and intestinal loop (35 cm) with a number of 9 lymph nodes excised . To the histopathology, the resected specimen appeared encapsulated and translucent with hemorrhagic areas, showing nodular surface, and had a maximum diameter of 4 cm. At the immune-morphologic analysis, S-100 protein expression was positive, C-Kit expression was negative. All these features tended to classify the specimen as a so-called “ancient schwannoma”. Resection margins and 9 lymph nodes excised were disease-free. The patient was discharged after 5 Days.

3. Discussion

Schwannomas (or neurilemmomas) were first described by Verocay in 1908[2]. They are generally solitary benign tumors originating from the Schwann cells of the peripheral nerves sheath, inclosing the sympathetic and parasympathetic nervous fibers[1].Frequent localizations are the head (intracranial tumors arise most often from the eighth cranial nerve)[3,4] and neck regions (particularly in the parapharyngeal space)[5,6], and also on the flexor surfaces of the extremities[7]. According to some authors[8], they tend to affect women more often than men and are seen in the third to the sixth decade of life[9]; some others[10] assert that they may be found in all age groups, butmore commonly in the first four decades and affect both sexes equally. The exact incidence of schwannomas is unknown, but they are rare[10] and arise in association with neurofibromatosis type I (NF-1) or develop sporadically. Anyway, reliable epidemiological data could not be extrapolated from such a poor information found in literature, mostly due to the rarity of the lesion and lack of major studies. Malignant transformation is overly rare and it frequently seems to be in association with Von Recklinghausen’s disease (NF-1). For their histopathological features, these lesions are frequently thought to be misdiagnosed neurofibromas[11]. Nevertheless, between 2% to 10% of head and neck schwannomas are malignant[7]. They are not generally associated with pain or neurologic symptoms, except when they occur in the setting of Von Recklinghausen’s disease. Any pain or neurologic symptom is usually associated to mass increase or malignant changes. Indeed, patients with a malignant change in schwannomas usually presented with rapid growing of a pre-existing lesion and pain.[12,13]For examples, tumors of the seventh nerve may be associated with facial paralysis[14];lesions involving the eighth nerve may cause giddiness or hearing loss[14];involvement of the ninth, tenth, and eleventh nerves may result with cough or dyspnea[14];tumors of the twelfth nerve may determine tongue impairment[14];lesions of the fifth nerve are usually asymptomatic[14]. Schwannomas developing within the bone are infrequent, generally affecting the mandible, and causing swelling, pain, or paresthesia[15]. They are well-defined, unilocular radiolucencies[16] that may be misdiagnosed with primordial

cysts, dentigerous cysts, or ameloblastomas[17]. They may determine local bony destruction and can be confused clinically with more malignant lesions[18]. This predilection is understandable because no other bone contains a canal that transmits a neurovascular bundle of such length and size.[18,19] In the soft tissue, they grow up as single, sessile or firm, rubbery, circumscribed nodules[14,20,21]. The larger tumors appear irregularly lobulated and may be partly cystic[21]. Generally, encapsulated lesions are freely mobile except for some rarest pedunculated or dome-like variety[22, 23]. Regarding the histopathology, these lesions usually consist of a cellular component (Antoni type A area) and a loose hypocellular component (Antoni type B)[10]. Antoni A areas are made of compacted tapering cells generally organized in palisades or in organoid clusters (Verocay bodies). Depending on the histopathology, several variants have been observed, such as cellular, glandular and epithelioid, and all generally seem to have a benign progression[10]. Cellular schwannomas are almost totally characterized of Antoni A areas without Verocay bodies[10].The cases of inveterate schwannomas showing central degenerative changes with cyst formation, calcification, hemorrhage, hyalinization and xanthomatous infiltration as demonstrated in the described case here are defined ancient schwannomas[24]. Ancient schwannoma is a rare variant. It was first characterized by Ackerman and Taylor in 1951[25,26]. They generally occur in the head and neck region[27] (trigeminal nerve, facial nerve, vestibular nerve, vague nerve, parotid, thyroid, vocal cord, floor of mouth, orbit and infra-temporal fossa). Other rare sites are the extremities, mediastinum, thorax[28] retroperitoneum[29], pancreas[30] and pelvis[31]. Furthermore, even scrotal schwannomas have been infrequently described in the scientific literature[32,33,34].They show singular hyperchromatic nuclei without exceeding mitoses.Ancient schwannoma shows benign degenerative histological features occurring over time[22,35]. These changes have been found to be a relative frequent phenomenon and include cystic, myxoid, edematous and fibrotic areas, vascular abnormalities, calcifications and atypical cells with pleomorphic nuclei[36]. Some authors believe that a tumor size increase could determine a central vascular failure: this could be the cause of central degenerative changes[37]. Ancient schwannoma is thought to be much more like a benign neural neoplasm, and the cytologic atypia seen in this tumor should be considered as a reactive/regressive phenomenon, not certainly as an evidence of malignancy[36,38]. Anyway, before pathologists realized that hypercellularity and atypia were not malignant features, many of these lesions were wrongly diagnosed as sarcomas[39]. Dahl (1977)[40] reported 6 out of 11 cases of ancient schwannoma to have been originally misdiagnosed as a malignant tumor (such as sarcoma)[41]. Moreover, a high expression of Antoni type B tissue and the loose architecture with regression areas caused by vascular thrombosis, gives the lesion a pseudocystic appearance in the CT scan so that it may lead to an erroneous diagnosis of epithelial tumors (microcystic serous adenoma, solid and papillary neoplasm and mucinous macrocystic tumor)[42]. Clinically and histologically, schwannoma should be differentiated from neurofibroma, which is a more common neurogenic tumor consisting of a mixture of Schwann cells and fibroblasts[43]. Neurofibromas are not encapsulated, and, unlike the neurilemmoma, the associated nerve becomes entangled with the neoplasm[44,45].It appears more frequently and often presents in a multiple fashion and in conjunction with NF-1[44,45].Because neurofibroma are more frequently associated with malignant transformation, it is importance to differentiate it from schwannoma, probably because Schwann cells are much more differentiated and don't seem to tend to neoplastic proliferation[43,45,17].The differential diagnosis also comprises primary benign or malignant soft tissue lesions, as well as malignant lymphoma. Furthermore, secondary tumors should be recognized[1].Thus, schwannomas are not certainly easy to diagnose. Head and neck schwannomas often put pathologists into confusion and preoperative investigations are frequently unavailing[46]. Radio-pathological features are often non-specific[47]. Ultrasonography can differentiate between solid and cystic tumors. CT can be helpful in determining the size, location, local involvement and distant spread[10]. Magnetic resonance imaging (MRI) could provide better visualization of the tumor. Fine-needle aspiration biopsy gives disputable results[48]. Fine needle aspirate (FNA) cytology is not really helpful because cytological specimens don't seem to show the exact architectural structure of the lesion[29], and have the potential to confuse this lesion with a more serious one[49]. In the current case, the diagnosis was only evident post-operatively. The only gold standard diagnostic investigation still remain histology after total excised specimen[41]. These lesions should be treated by total surgical excision (with every surgical effort made to preserve nerve integrity). If the nerve is too thin or splayed out, grafting of the defective nerve segment should be the appropriate approach[39]. Although benign, large and incompletely excised lesions are capable of recurrence[28,34].

4. Conclusion

Primary benign schwannoma of the mesentery is extremely rare with less than ten previously described cases. the clinical signs and symptoms of ancient schwannoma tend to be non-specific or even absent; they are difficult to diagnose with cross-sectional imaging and histopathological examination is required to reach a final diagnosis. This entity should be considered in the differential diagnosis of slow-growing abdominal masses.

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