OLGU SUNUMU CASE REPORT

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Malignant Presentation of Benign Tumour of the Rectum

Rektumun Benign Tümörünün Malign Görüntüsü

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ABSTRACT Rectal schwannomas are rare mesenchymal tumour of the gastrointestinal tract. They are scarcely identified in the low rectum as compared to the proximal tract. If present, huge tumour tends to be symptomatic leading to obstruction, bleeding, and tenesmus. We highlight a 74-year-old man with malignant presentations mimicking low rectal cancer. Colonoscopy revealed a huge pedunculated polyp measuring 10 x 8 cm with a diagnosis of benign tumour of schwannoma upon punch biopsy. Transanal excision was made after failure of endoscopic resection. Microscopic evaluation of the tumour showed a benign nerve sheath tumour of schwannoma supported by positive S100 protein expression. His recovery was excellent, hence he was discharged on the following day. There are no similar symptoms and tumour recurrence on follow up. A huge rectal tumour is not always malignant. Despite their rarity, schwannomas can manifest as malignant features. In case of tumour that mimics rectal carcinoma, preoperative diagnosis is very important for a management plan. The definitive histological diagnosis is based on the morphological features with diffuse positivity of S100 from immunohistochemical study. Surgical resection either endoscopic or transanal approach is the mainstay of treatment, especially when dealing with local complication caused by the huge bulk of tumour.

Keywords: Schwannoma; rectum; colonoscopy

ÖZET Rektal schwannoma gastrointestial sistemin nadir görülen mezenkimal tümörüdür. Proksimal kısma göre alt rektal bölgede nadiren görülür. Dev tümörler semptomatik olma eğiliminde olup obstrüksiyon, kanama ve tenesmusa yol açabilir. Bu olgu sunumunda alt rektal kanseri taklit eden malign presentasyonlu 74 yaşında bir erkek hastayı bildirdik. Kolonoskopide 10x8 cm boyutlarında saplı dev polip görülmüş olup punch biyopsi sonucu schwannoma tanısı koyuldu. Endoskopik rezeksiyon başarılı olmadığı için transanal eksizyon yapıldı. Tümörün mikroskopik değerlendirilmesi sonucunda benign sinir kılıfı tümörü schwannoma tanısı S100 protein ekspresyonu ile desteklendi. Hasta mükemmel şekilde iyileşti ve ertesi gün taburcu edildi. İzlemde benzer semptomlar ve tümör rekürrensi görülmedi. Dev bir rektal tümör her zaman malign değildir. Schwannomlar nadir görülmesine rağmen malign özellikler sergileyebilir. Rektal kanseri taklit eden tümör varlığında tedavi planı için preoperatif tanı çok önemlidir. Kesin histolojik tanı immün histokimyasal çalışmada diffüz S100 pozitifliği ile morfolojik özelliklere dayanır. Endoskopik veya transanal yaklaşımla cerrahi rezeksiyon, özellikle de tümörün dev boyutunun neden olduğu lokal komplikasyonlarla mücadele ederken tedavinin esasını oluşturur.

Anahtar Kelimeler: Schwannoma; rektum; kolonoskopi

Schwannoma is an abnormal growth that originates from Schwann cells, which originally form the neural sheet and encompass the nerve of myenteric plexus. Review of literature has suggested that gastrointestinal schwannoma accounts for 2-8% of all mesenchymal tumours frequently found in stomach and small intestine, while its location in colon and rectum is scarce.[1] Rectal schwannomas are exceptional with only a few cases have been reported.[2] Mostly are asymptomatic and only discovered dur-

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ing screening endoscopy or incidentally during abdominal imaging. However, a huge tumour size tends to cause symptoms such as obstruction, bleeding, and tenesmus. The diagnosis is confirmed by morphology supported by immunohistochemical panel of S100 protein. Excision with a clear margin is mandatory due to their tendency to recur locally or become malignant if it's left untreated.[3] We describe a case of benign tumour of rectal schwannoma in a 74-year-old male, who presented with alarming symptoms masquerading as rectal carcinoma, which was successfully treated by transanal excision of the tumour.

CASE REPORT

A 74-year-old male was referred from a district hospital for per rectal bleeding and tenesmus with a palpable mass on rectal examination. The mass was felt 4 cm from the anal verge which intrigued us for further management. Despite lacking family history of malignancy, he presented with altered bowel habits, tenesmus, mucus discharge and per rectal bleeding for 3 months. Colonoscopy revealed a malignant looking pedunculated polyp 4 cm from the anal verge and the stalk origin was 8 cm from the anal verge. A punch biopsy was consistent with benign tumour of schwannoma. A traditional colonoscopic snaring of the polyp was unsuccessful. He then underwent a transanal excision of the rectal polyp. Intraoperative finding showed a large 10x8 cm anal polyp, hard in consistency with stalk 10 cm from the anal verge.

An excision was made at the stalk and it was sent as a whole for histopathological examination. Macroscopic appearance revealed a massive rounded mass measuring 10 x 10 cm with the smooth mucosal surface (Figure 1). The microscopic view showed Antoni A and Antoni B areas of spindle-shaped cells with presence of Verocay bodies, supported by positive S100 protein and vimentin (Figure 2). They were negative for CD34, CD117, desmin and actin. There was no proper capsule identified as the resected specimen was ulcerated. However, histologically the tumour was well defined and non-infiltrative. There was no evidence of malignant transformation. He recovered well postoperatively and was discharged home the following day. The final histopathological report was correlative to the preoperative diagnosis which was a benign tumour of schwannoma of the rectum.

DISCUSSION

Schwannomas are mostly benign (90%) and often arise in the head and neck (25-40%) but not in the retroperitoneum, colon and rectum (1%).[4] Although rare, it should be considered to be the differential diagnoses of slow-growing tumours in anal region such as lipomas, leiomyosarcomas, neurofibromas, ganglioneuromas, paragangliomas, granular cell tumours, and glomus tumours. Gastrointestinal schwannomas occur slightly more in female patients (almost 60%) with a mean age of 61.5 years.[1] They can appear as a submucosal lesion, mucosal mass or an even pedunculated polyp as in our case.

Despite having a benign histology, a huge tumour size in the anal region can be symptomatic masquerading as features of a malignant tumour. The patient can manifest false alarming but yet crucial presentations namely per rectal bleeding (from tumour ulceration), tenesmus and mucus discharge as well as colonic obstruction. Majority of the presenting complaints from our patient mimic a red alert of low rectal cancer, in which an urgent intervention provides a meaningful outcome. However, not every alarming symptom is proven to be menacing. A preoperative clinical diagnosis is important hence biopsy of tumour and histopathological examination plays a pivotal role in surgical approach for a patient with tumour at the anal region. The findings of Antoni A and Antoni B areas of spindle-shaped cells with the presence of Verocay bodies are pathognomonic of schwannoma. Since they are neurogenic tumours,

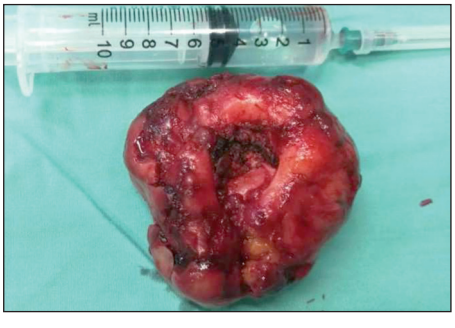

FIGURE 1: Macroscopic specimen after transanal excision of the rectal polyp. Intraoperative finding showed a large 10 x 8 cm anal polyp.

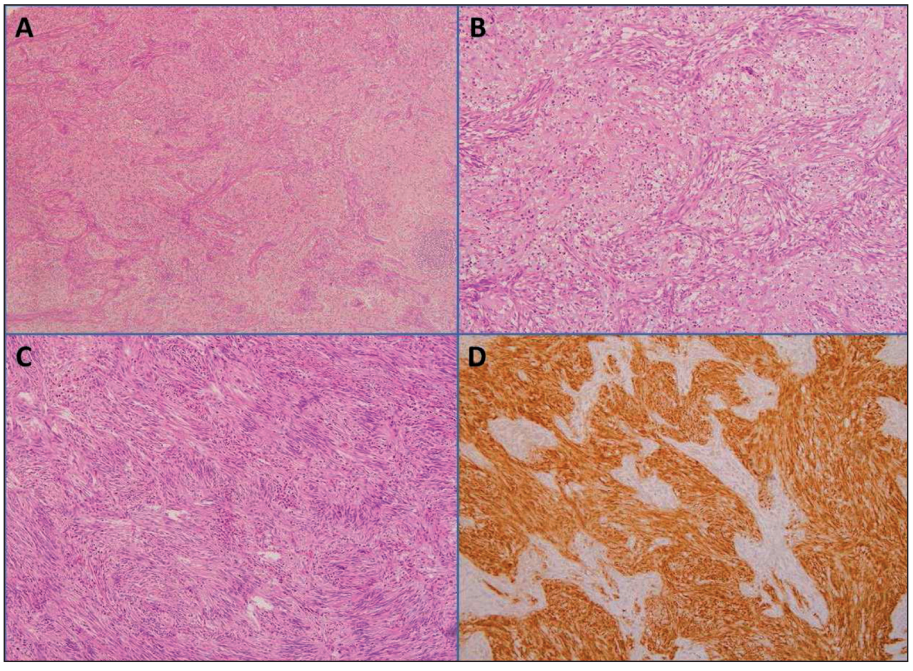

FIGURE 2: (A) Histopathological evaluation revealed a biphasic pattern of tumour cells composed of hypercellular Antoni A and hypocellular Antoni B areas. (Hematoxylin and eosin (H&E), original magnification x4). **(B)** Antoni B myxoid hypocellular areas with scattered inflammatory cells infiltrate. (H&E, original magnification x20). **(C)** Anthony A areas showing the tumour cells are of elongated, wavy spindle-shaped cells with tapered ends interspersed with collagen fibers. Note the presence of nuclear palisading forming Verocay bodies. (H&E, original magnification x10). **(D)** Immunohistochemical stain showing a strong, diffuse staining of S100. (Immunohistochemistry, original magnification x20).

they are usually positive for S100 protein and vimentin while negative for desmin, keratin, glial fibrillary acidic protein, CD34, EMA, smooth muscle and muscle-specific actin.[5]

Among the differential diagnoses that might be considered in the case of schwannoma are neurofibromatosis, schwannomatosis and melanocytic tumours. The rectal polyp was the only mass that was identified. There were no other symptoms and signs of neurofibromatosis type 1, neurofibromatosis type 2 or schwannomatosis noticed.[6] In this case, there were no features of melanocytic tumours clinically, grossly and microscopically as the resected specimen showed a typical histological feature of schwannoma.

Since this is proven to be rectal schwannoma, a decision for surgical excision was made. In general, surgical approach in rectal tumour depends on the size, location and histopathological pattern of the tumour.[7] Endoscopic and minimally invasive approaches are still proven to be beneficial. In our case, transanal excision of the rectal polyp was the best approach in view of the immensity of the tumour.

In conclusion, schwannomas of the gastrointestinal tract are rare especially in the rectum. Preoperative histological diagnosis is paramount, especially when dealing with tumour mimicking adenocarcinoma. The definitive diagnosis is based on classical histology features, assisted by immunohistochemistry of S100 protein. Surgical resection is the mainstay of treatment notably concerning with local complication caused by the huge bulk of tumour.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

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