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CLINICAL CASE

Pleomorphic sarcoma of the jejunum: Case report

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Abstract

Small bowel tumors are rare and account for 1-2% of all gastrointestinal neoplasms and < 5% of all malignancies in the gastrointestinal tract. A 65-year-old man presented with anemia and melena for approximately 6 months, treated with several blood transfusions. Endoscopy is performed, finding gastritis atrophy. The patient underwent contrast computerized tomography identifying a tumor in jejunum proximal approximately 3 cm diameter, so laparotomy is performed finding tumor from 30 cm of Treitz angle, and it was performed resection with clear margins. Histological examination revealed pleomorphic sarcoma of jejunum.

Keywords: *Pleomorphic sarcoma. Jejunum. Small intestine.*

Sarcoma pleomórfico del yeyuno: reporte de caso

Resumen

Los tumores de intestino delgado son raros, constituyen el 1-2% de todos los tumores de tubo digestivo y menos del 5% son malignos. Se presenta el caso de un hombre de 65 años de edad que presenta anemia y melena de seis meses de evolución, tratado con transfusiones sanguíneas. Se realizan estudios de endoscopia, identificando gastritis atrófica. Se complementa con tomografía computarizada, la cual identifica un tumor en yeyuno proximal de 3 cm diámetro. Se interviene quirúrgicamente, identificando tumor a 30 cm de ángulo de Treitz y se reseca con márgenes libres. Resultado por inmunohistoquímica: sarcoma pleomórfico de yeyuno.

Palabras clave: *Sarcoma pleomórfico. Yeyuno. Intestino delgado.*

Introduction

Small bowel malignant tumors are very rare, with an annual incidence of 22.7 per million and the sarcomas rank fifth (1.2%) among small bowel malignant tumors[1]. Management of these tumors is a challenge because of their rarity, relative inaccessibility for diagnosis, various histologic types, and non-specific symptoms[2]. These sarcomas arise in distal extremities, they are a frequent form of sarcoma in the elderly and tend to spread to lung[3]. The most frequent clinical presentation of jejunoileal tumors includes: recurrent abdominal pain, abdominal mass, intestinal obstruction or perforation, and gastrointestinal hemorrhage[4].

Case presentation

A man 65 years old admitted to the emergency department with history of fatigue, melena, anemia, and

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abdominal pain. Physical examination showed no abdominal mass and skin pallor. Routine laboratory showed just anemia (8.7 g/dl) normochromic, and normocytic, no abnormalities were observed in the rest of studies.

Suspecting bleeding in the upper gastrointestinal tract a gastroscopy was performed revealing gastropathy in the body and antrum, a colonoscopy also showed diverticulum in the right colon and hemorrhoidal disease.

These findings could not explain the cause of anemia, later, hemoglobin was decreased until 5 g/dl a computed axial tomography was performed with administration of intravenous contrast, so a 3 cm tumor was identified in the proximal jejunum, the rest of tomography did not show any local or distant metastases. Laparotomy is performed and they found a tumor originating from the wall of the small bowel (jejunum) no sites of abdominal dissemination were identified and wide resection of the tumor with end to end anastomosis was performed.

Pathological examination of the specimen revealed tumor of 5 × 3 cm, malignant mesenchymal tumor, free proximal, and distal margins. About immunohistochemistry, cells negative to pankeratin ae1/ae3, S100, CD117, CD34, and CDK4. Final diagnosis concluded pleomorphic sarcoma of jejunum.

The patient evolves successfully without complications and left hospital after 10 days of surgery. He kept on regular follow-up by 12 months. His last clinical examination and abdominal tomography showed no evidence of relapse.

Discussion

Small bowel tumor is very rare, accounting for 1-2% of gastrointestinal neoplasm[5]. Malignant soft-tissue tumors of small intestine are extremely rare, the most common type is leiomyosarcoma[6]. The malignant small bowel tumors are only 2%, adult sarcomas are located in the gastrointestinal system[7].

Undifferentiated pleomorphic sarcoma (UPS), which was formerly known as malignant fibrous histiocytoma, is a high-grade sarcoma, which mainly arises from the soft tissue of the extremities and can appear at any age[8]. UPS is the most common type of high-grade malignant sarcoma found in elderly people and most are asymptomatic[8].

Primary benign or malignant tumors of the jejunum and ileum are even more rare, intestinal obstruction accounts from 12% to over 50% clinical presentations of jejunoileal tumors[9].

Intestinal obstruction accounts from 12% to over 50% clinical presentations of jejunoileal tumors, nine small bowel tumors often go untreated for a long time because they are a great challenge to diagnosis and treatment[9].

Jejunoileal tumors are suspected only under extreme conditions such as the presence of a palpable abdominal mass, gastrointestinal hemorrhage, intestinal perforation, or obstruction[9]. Our case was treated with laparotomy for the history of anemia and findings of tumor in the tomography after negative findings by gastroscopy and colonoscopy.

Although there are no specific findings by computerized tomography for pleomorphic sarcoma as far as we know, the previous review of peritoneal sarcomatosis reported that peritoneal implants and mesenteric involvement were well-defined and neither diffuse thickening nor calcifications were asociated[9].

For intestinal obstruction secondary to jejunoileal tumors, resection of the obstructed segment may afford the best outcome[9]. Emergency surgery for these patients precludes a complete and negative margin resection and constitutes a risk factor for residual disease and short-term survival[9].

About sarcomas retrospective studies determined that an adequate resection (wide margins) during treatment is the single most important prognostic factor for long-term survival as local relapse and distant metastases[10].

The role of adjuvant radiotherapy is well established for treating pleomorphic sarcoma of the extremities but the effects of radiotherapy for treating patients with mesenteric pleomorphic sarcoma remain unclear, and chemotherapy is reserved for patients with unresectable or metastatic tumors[11].

Diagnosing pleomorphic sarcoma requires to exclude other kind of tumors such as gastrointestinal stromal tumors, pleomorphic liposarcoma, pleomorphic leiomyosarcoma, pleomorphic rhabdomyosarcoma, dedifferentiated liposarcoma, myxofibrosarcoma, poorly differentiated carcinoma, and melanoma[11].

In our case, the diagnosis was confirmed with immunohistochemical staining demonstrated for negative to pankeratin ae1/ae3, S100, CD117, CD34, and CDK4. The patient kept on regular follow-up by 12 months. His last clinical examination and abdominal tomography showed no evidence of relapse.

Conclusion

Pleomorphic sarcoma of the small intestine is a rare neoplasm. Diagnosis is corroborated for immunohistochemical staining and, the treatment of these tumors is

the resection with free margin, actually without evidence about the benefit of adjuvant chemotherapy or radiotherapy.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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