

Colonic Infiltration By A Metastatic Malignant Peripheral Nerve Sheet Tumor Arising In The Lower Limb

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Citation

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Abstract

A 74-year-old man presented a mass located in the left leg near the knee which was characterized as a malignant peripheral nerve sheet tumor (MPNST). Multiple lung lesions and a 5-cm mass in the caecum were detected by Computed Tomography. This appearance was equivocal. By colonoscopy and related biopsies colonic infiltration by sarcoma cells was detected.

In this case, a primitive MPNST and bowel metastasis were diagnosed. Given the relative rare tumor it is difficult to estimate the incidence of bowel metastasis, but secondary bowel involvement in MPNST must be taken into consideration in presence of suggestive radiographic images and/or clinical symptoms (bleeding and anemia).

CASE REPORT

The patient, a 74-year-old man, presented at the observation with a mass located in the right leg near the knee measuring about 10 cm. In the inguinal right region there were two lymphonodal packets measuring, respectively, 6 cm and 4 cm in their largest diameter. The general state of health was good. At the biochemical examination, the patient presented neutrophil leukocytosis (WBC $32.66 \times 10^3 / \mu\text{L}$, Neutrophils $28.32 \times 10^3 / \mu\text{L}$), a mild microcytic anemia (12 g/dL), ESR 35 mm/h (normal range: 1-10), Alkaline Phosphatase 169 U/L (normal value: <122). The mass in the leg, characterized as a malignant peripheral nerve sheet tumor (MPNST), grew rapidly and bled. Computed Tomography showed multiple nodular lung lesions, more concentrated in lower lobes, and a 5-cm mass in the caecum, which did not take contrast homogeneously (fig.1).

Figure 1

Figure 1: Abdominal Computed Tomography: the arrow shows a 5-cm mass in the caecum.

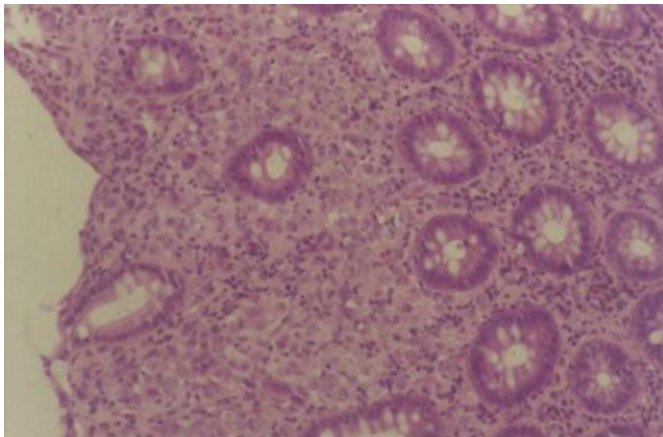


This appearance was equivocal. The patient did not present either signs or symptoms related to probable bowel disorders. By colonoscopy one sessile and two polyps were detected respectively in the right colon and in the caecum, and neither infiltrating masses nor stenosis were showed. For the rest, mucosa appeared uniformly rosy without signs of

recent bleeding. Infiltration of soft tissue tumor cells in the mucosa was revealed in the colon sample (fig.2), while no tumor cells were documented in the tissue removed from the caecum. The metastasis was formed by both clustered and scattered tumor cells and was confined in the mucosa.

Figure 2

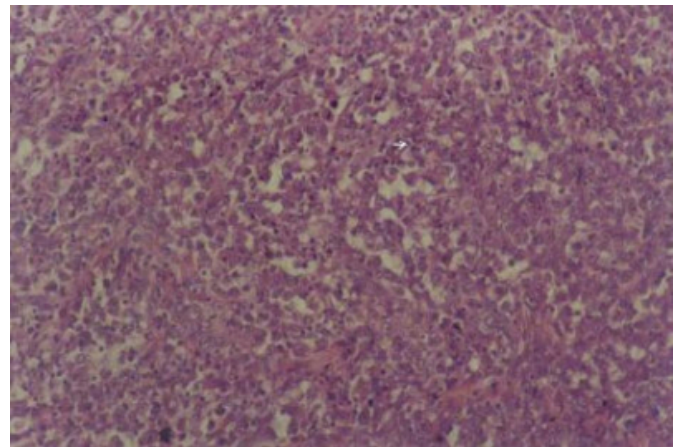
Figure 2: Periodic Acid Schiff stain, 10× magnification: soft tissue tumor infiltration of colonic mucosa. Nuclei changes can be visualized such as deep strangulation indicated by the arrow.



The patient was submitted to amputation of lower right limb because of complicated overlapping infection. Epithelioid MPNST was confirmed (fig.3). Neoplastic cells showed high mitotic index and vascular invasion. Skeletal muscle and skin were both infiltrated, while bone and surgical lips were uninjured. Immunohistochemically, the tumor was shown to express vimentin and neuron specific enolase. S100 and AE1-AE3 cytokeratins were expressed focally and desmin, smooth muscle actin and HMB-45 were not expressed. Inguinal right lymphnode was also confirmed to be infiltrated by cancer cells.

Figure 3

Figure 3. Hematoxylin-eosin stain, 4× magnification: Primitive soft tissue tumor lesion. Epithelioid cells in mitosis or binuclear can be recognized, as indicated by the arrow.



Case reports (¹) and a small series (²) of gastrointestinal primitive schwannomas report a more frequent growth in the caecum and rectosigmoid colon. Presenting symptoms typically drawing the attention on gastrointestinal tract are described, i.e. rectal bleeding, colonic obstruction and abdominal pain. The tumors range from 0.5 to 5.5 cm in diameter and have a low mitotic index. They do not present together with neurofibromatosis and behave in a benign fashion without recurrence or metastasis.

Conversely, in the case previously described, bowel infiltration by sarcoma was diagnosed. Intestinal metastases by bone or visceral and limb soft tissue tumor are extremely rare findings (^{3,4,5,6,7,8,9,10,11,12,13,14}). Gastrointestinal metastases by musculoskeletal sarcomas were reported in 6 out of 505 patients (1.2%) (¹⁵). In most cases clinical symptoms, bleeding (^{3,8}), obstruction (^{5,6,10,11,12,13}) or perforation (⁹), were present. To our knowledge only one previous case with similar histology, a left leg malignant schwannoma, was reported in literature (¹⁶). Differently from our patient, a progressive anemization without evidence of bleeding led to the identification of an ulcerative lesion at the ileocolonic junction (¹⁶). The two cases together seem to suggest a predilection for the right colon.

Mesenteric and peritoneal metastases frequently occur through hematogenous and lymphatic patterns (¹⁷). Metastasis within bowel mucosa are very rare events. Polypoid appearance of metastasis similar to that described in the presented case was, however, common to other reports (^{4,8}). Nevertheless, larger masses (⁴) or multiple hemorrhagic polypoid lesions (⁸) were described, while in the presented

case what appeared as a polyp was really a lifting of mucosa with below infiltration by tumor cells.

Soft tissue tumors usually metastasize to the lungs and to the liver. Lungs are the only sites of metastatic disease in approximately one half of the patients. Extrapulmonary metastases can occur as a late manifestation of widely disseminated disease. In this setting the involvement of large bowel by soft tissue tumors with neurogenic differentiation is surely an out-of-ordinary pattern of metastatic spread.

In conclusion, the secondary bowel involvement in MPNST described in the present case is an atypical site of metastatization, which could be often unrecognized, but must be considered in presence of suggestive radiographic images and/or clinical symptoms (bleeding and anemia).

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References

1. Hirose T, Scheithauer BW, Sano T. Giant plexiform schwannoma: a report of two cases with soft tissue and visceral involvement. *Mod Pathol* 1997;10:1075-81.
2. Miettinen M, Shekitka KM, Sobin LH. Schwannomas in the colon and rectum: a clinicopathologic and immunohistochemical study of 20 cases. *Am J Surg Pathol*. 2001; 25:846-55.
3. Tamura M, Oda M, Ohta Y, Omura K, Nonomura A, Watanabe G. Small intestinal bleeding secondary to metastatic malignant fibrous histiocytoma of the rib: report of a case. *Surgery Today* 2002; 32:69-71.
4. Panizo-Santos A, Sola I, Lozano M, de Alava E, Pardo J. Metastatic osteosarcoma presenting as a small-bowel polyp. A case report and review of the literature. *Arch Pathol Lab Med* 2000; 124: 1682-4.
5. Hung G-Y, Chiou T-J, Hsieh Y-L, Yang M-H, Chen WY-K. Intestinal metastasis causing intussusception in a patient treated for osteosarcoma with history of multiple metastases: a case report. *Jpn J Clin Oncol* 2001; 31: 165-7.
6. Ganguli SN, Hamilton P, Hanna S, Morava-Protzner I. Small bowel intussusception secondary to osteogenic sarcoma metastasis: case report. *Can Assoc Radiol J* 1999; 50: 170-2.
7. Costa A, Fuscaino L, Mosca F. Metastatic osteosarcoma involving the colon and ileum. *Gastrointest Endosc* 2001; 54:75.
8. Bandorski D, Arps H, Jaspersen D, Diehl KL. Severe intestinal bleeding caused by intestinal metastases of a primary angiosarcoma of the thyroid gland. *Z Gastroenterol* 2002; 40: 811-4.
9. Ise N, Kotanagi H, Morii M et al. Small bowel perforation caused by metastasis from an extra-abdominal malignancy: report of three cases. *Surg Today* 2001; 31:358-62.
10. Gerst PH, Levy J, Swaminathan K, Kshetry V, Albu E. Metastatic leiomyosarcoma of the uterus: unusual presentation of a case with late endobronchial and small bowel metastases. *Gynecol Oncol* 1993; 49: 271-5.
11. Avagnina A, Elsner B, De Marco L, Bracco AN, Nazar J, Pavlovsky H. Pulmonary rhabdomyosarcoma with isolated small bowel metastasis. A report of a case with immunohistochemical and ultrastructural studies. *Cancer* 1984; 53: 1948-51.
12. Kanoh T, Shirai Y, Wakai T, Hatekeyama K. Malignant fibrous histiocytoma metastases to the small intestine and colon presenting as an intussusception. *Am J Gastroenterol* 1998; 93: 2594-5.
13. Gorman RC, Jardines L, Brooks JJ, Daly JM. Enteroenteric intussusception due to a metastatic malignant fibrous histiocytoma. *J Surg Oncol* 1993; 54: 203-5.
14. Sondenaa K, Soreide JA, Erichsen C et al. Metastases in the small intestine from a subcutaneous lower limb leiomyosarcoma. *Acta Oncol* 1992; 31: 865-6.
15. Ogoe A, Morita T, Hotta T, et al. Intra-abdominal metastases in musculoskeletal sarcomas. *J Orthop Sci* 2000; 5:463-9.
16. Crespi C, Origgi C, Zampieri A, Calandra C, Crivelli F. Multiple jejuno-ileal metastasis of malignant schwannoma from the leg. Report of a case presenting anemia symptoms and review of the literature. *Minerva Chir* 1996; 51:829-33.
17. Carmignani CP, Sugarbaker TA, Bromley CM, Sugarbaker PH. Intraperitoneal cancer dissemination: mechanisms of the patterns of spread. *Cancer Metastasis Rev.* 2003 Dec;22:465-72.

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