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Superior Vena Cava Syndrome: An Uncommon Presentation of a Rare Colon Carcinoma Metastasis

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ABSTRACT

Introduction: Superior vena cava (SVC) syndrome is caused by obstruction of the superior vena cava due to vascular compression by a mass or intrinsic obstruction. The authors describe SVC syndrome caused by an isolated metastatic mediastinal mass from a resected primary colon carcinoma.

Case Report: An 81-year-old woman was referred to the hospital with swelling of the neck and upper left limb, dysphonia and dysphagia, associated with an involuntary weight loss of 16 kg. Mediastinal metastasis of colon adenocarcinoma was found, causing the SVC syndrome. The mass was unresectable and the patient was referred to palliative radiotherapy.

Discussion: Only 12 cases of mediastinal metastasis from colorectal cancer have been reported in the English literature.

Conclusion: As a rare manifestation of colorectal cancer, the presented case highlights the need for clinicians to be aware of rare metastases at the time of diagnosis.

LEARNING POINTS

- Superior vena cava (SVC) syndrome can result from vascular compression by a mass.
- Although mediastinal lymph node metastasis is rare in colorectal cancer, physicians should be aware of less common locations.
- Patients should have a close follow-up in order to avoid the growth of unresectable metastases, since surgery, when possible, can lead to a better prognosis.

KEYWORDS

Colon cancer, metastasis, mediastinal mass, subcutaneous nodules

CASE REPORT

We describe the case of an 81-year-old Caucasian woman who was referred to our hospital with swelling of the neck and upper left limb and a 2-week history of dysphonia and dysphagia, associated with an involuntary weight loss of 16 kg over the previous 2 months. Ten months earlier she had been diagnosed with ascending colon adenocarcinoma stage IIIb without distant metastatic disease (T4aN1b.IVL), and underwent right hemicolectomy. She was not eligible for adjuvant chemotherapy as she had stage 3 chronic kidney disease and ischaemic cardiomyopathy. On physical examination, she had face and neck oedema with visible collateral veins. She had three abdominal non-painful subcutaneous nodules, but no further relevant abnormalities.

Laboratory tests showed anaemia (haemoglobin 10.5 g/dl; normal range 12.0–16.0 g/dl), a normal serum carcinoembryonic antigen (CEA, 5 ng/ml; normal range 0–5 ng/dl) and elevated cancer antigen 19-9 (57 U/ml; normal range <37 U/ml). A chest x-ray revealed enlargement of the mediastinum (*Fig. 1*).

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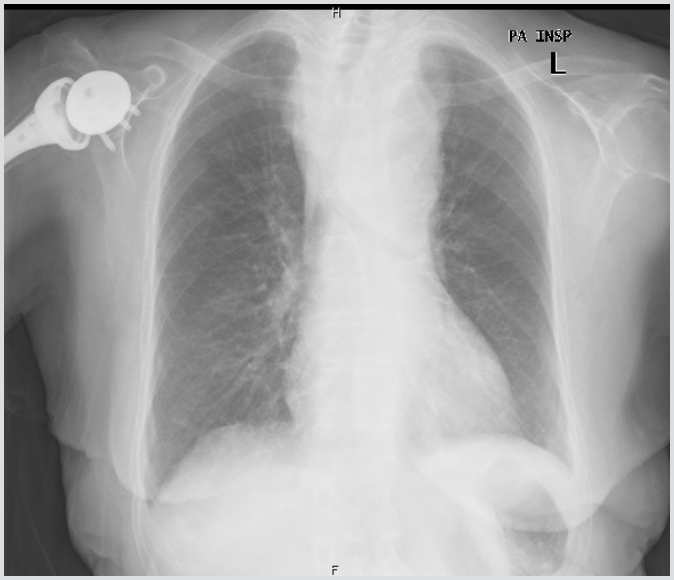

Figure 1. Chest X-ray showing an enlargement of the mediastinum

Therefore, upper endoscopy was performed and revealed an extrinsic compression area of approximately 4 cm (18–22 cm from the incisor teeth); no other changes were observed. An abdominal ultrasound only revealed three well-defined hypoechogenic subcutaneous nodules that could have been tumour implants. Computed tomography (CT) scans of the chest, abdomen and pelvis showed a large bulky solid tumoral lesion with heterogeneous density, measuring 9.0 cm at its greatest diameter, in the posterior mediastinum with invasion of the first two dorsal vertebral bodies, as well as of the supra-aortic trunks, close to the posterior wall of the trachea. It had invaded the oesophagus and the pleural space bilaterally (*Figs. 2–4*).

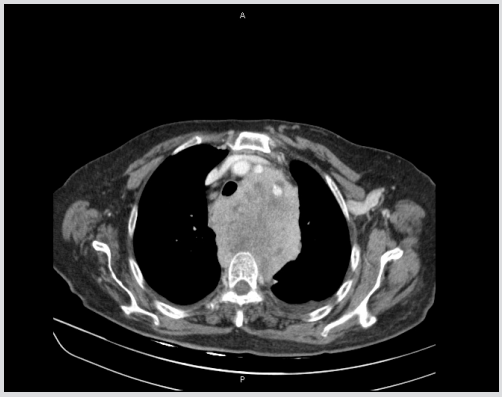

Figure 2. Computed tomography scan coronal cut revealing a large solid tumoral lesion in the posterior mediastinum

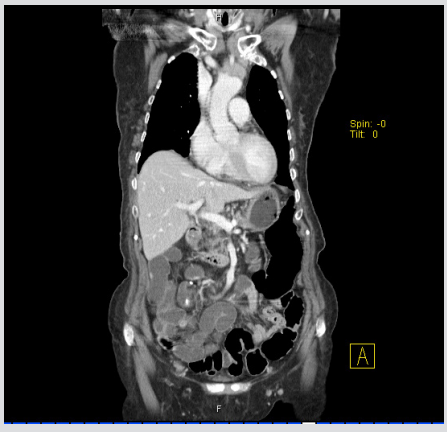

Figure 3. Computed tomography scan sagittal cut revealing a large solid tumoral lesion in the posterior mediastinum

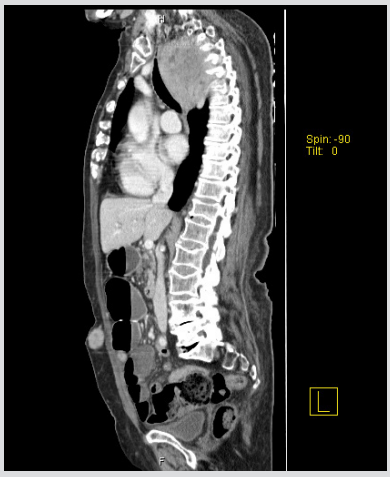

Figure 4. Computed tomography scan axial cut revealing a large solid tumoral lesion in the posterior mediastinum

Several cutaneous nodules on the abdomen compatible with metastases were found (*Fig. 5*). No metastases or other pathologies were identified in the lungs, and abdominal-pelvic CT images were unremarkable. CT-guided fine needle aspiration cytology of the mediastinal tumour and the subcutaneous nodules was compatible with metastatic adenocarcinoma. Immunohistochemical staining of the core biopsy samples was negative for cytokeratin 7 (CK7) and neuroendocrine markers, and positive for cytokeratin (CK) AE1/AE3, CDX2, CAM 5.2 and CK20, which is typical of colorectal primary carcinoma.

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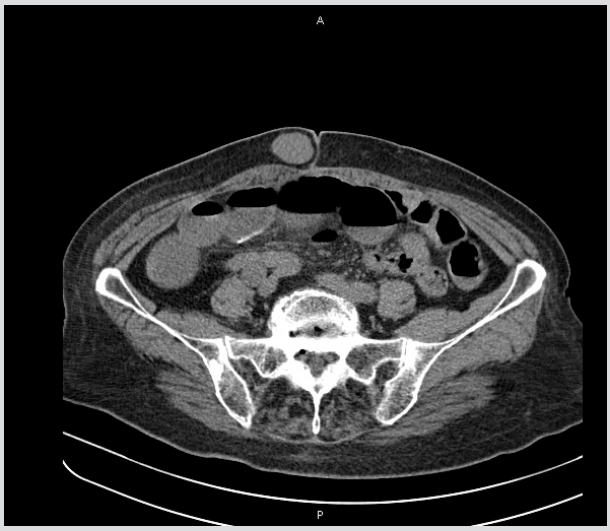

Figure 5. Computed tomography scan axial cut revealing cutaneous nodules on the abdomenlesion in the posterior mediastinum

DISCUSSION

This report describes a case of SVC syndrome that resulted from external compression by a metastatic mediastinal mass from colorectal cancer. This is a rare cause of SVC syndrome and itself an unexpected and rare metastasis. Over 80% of cases of SVC syndrome are due to malignancy, but generally from lung cancer or lymphoma[1]. Another uncommon finding in this case was the aetiology of the mediastinal mass, as the majority of lesions in the posterior mediastinum are lymphadenopathies, neurogenic tumours or cystic lesions. In addition, mediastinal lymph node metastases are rare in colorectal cancer, especially when there are no lung or liver metastases. A further peculiarity was the subcutaneous nodules, which are found only in 4–6.5% of metastatic colon adenocarcinoma cases[2]. As in our patient, the most common site is the abdominal skin and they are frequently related to poor prognosis.

Only 12 cases of mediastinal metastasis from colorectal cancer have been reported in the English literature (*Table 1*)[3–13]. However, this is the first reported case of a bulky isolated mediastinal mass with an exuberant presentation such a short time after primary tumour resection. In the majority of cases, the primary tumour was located on the ascending colon (41.6%) and 83.3% of patients were men. Although mediastinal metastasis is frequently related to liver or lung metastasis, only six cases presented with liver metastasis. Direct colon metastasis to the mediastinal lymph nodes was described in eight case reports. In nine cases, the mediastinal lymph node metastasis appeared more than 1 year after resection of the primary cancer. In one case, the metastasis led to the diagnosis of colorectal carcinoma as the primary cancer.

The pathway of this metastasis is unclear, but there are two possible routes: retrograde lymph node and haematogenous spread [14]. Kuba et al. proposed that the tumour cells from ovarian metastasis spread through the paravertebral venous plexus (remetastasis or metastasis of metastasis) or the para-aortic lymphatic drainage route[4]. Remetastasis of colon cancer to the mediastinal lymph nodes was also described from the lungs and liver. Vetto et al. reported a case of colon cancer metastasizing to the right lobe of the liver with an isolated mediastinal lymph node metastasis. The authors' hypothesis was that the lymphatics from the falciform ligament and right lateral lobe drained upwards through the diaphragm, caval foramen and oesophageal hiatus to the mediastinal nodes[3]. In fact, many cases of gastrointestinal cancer metastasize via the thoracic duct from a retroperitoneal lymph node (via retrograde lymph node metastasis). This may occur through incompetent valves of lymphatics as there is no communication between the thoracic duct and the bronchomediastinal trunk[15].

The diagnosis of a mediastinal mass is challenging as many of these masses have a similar appearance on imaging, varying only in certain details seen on CT. The location and composition of a mass together with the clinical history can lead to the diagnosis. In our case, despite expectations, the pathological diagnosis was conclusive and showed colon cancer metastasis[16]. The benefits of adjuvant chemotherapy are still uncertain, as is the role of radiotherapy in these patients. Surgery has been reported as a treatment for solitary lymph node metastasis, with a good prognosis. However, the efficacy of mediastinal metastasectomy is unproven because not enough cases have been described in the literature to support it. As mediastinal metastases of colon cancer are rare, the treatment strategy has yet to be established[17]. Although there are few reported cases of mediastinal metastasis from colorectal cancer, physicians should be aware of these less common locations and consider the possibility of mediastinal lymph node metastasis as a differential diagnosis. Patients should have a close follow-up in order to avoid the development of unresectable metastases, since surgery, when applicable, can lead to a better prognosis.

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Case number	Reference	Year published	Age	Sex	Primary location	Primary stage	Adjuvant chemotherapy	Time after Primary resection(months)	Metastasis location
1	Vetto et al. [3]	1991	60	M	Hepatic flexure	IIIb	Not listed	12	Right lobe liver, mediastinum
2	Kuba et al. [4]	1999	60	F	Sigmoid	IIIb	Not listed	33	Left ovary, mediastinum
3	Musullam et al. [5]	2008	67	M	Rectosigmoid	IIIb	Yes	26	Mediastinum
4	Yavaş et al. [6]	2009	57	M	Ascending	IIIa	5-Fluorouracil	Not listed	Mediastinum
5	Sano et al. [7]	2011	29	M	Ascending	IVa	5-Fluorouracil	24	Liver, mediastinum
6	Iwata et al. [8]	2012	75	M	Ascending and transverse	IIIa	Capecitabine	42	Liver, mediastinum
7	Matsuda et al. [9]	2014	65	M	Sigmoid	IIIc	Tegafur-uracil	101	Mediastinum
8	Matsuda et al. [9]	2014	50	M	Rectum	IIIc	No	96	Mediastinum
9	Halabi et al. [10]	2014	44	M	Ascending	IIIa	Folinic acid, fluorouracil, oxaliplatin, bevacizumab	22	Mediastinum
10	Shirakawa et al. [11]	2015	65	M	Rectum	IIIa	Tegafur-uracil, 5-fluorouracil, leucovorin and oxaliplatin	55	Liver, mediastinum
11	Rodríguez-López et al. [12]	2016	45	M	Rectum	IVa	Folinic acid fluorouracil and oxaliplatin, bevacizumab	–	Mediastinum
12	Toda et al. [13]	2017	59	M	Ascending	IIIb	Yes	32	Mediastinum

Table 1. Cases of mediastinal metastasis from colorectal cancer reported in the English literature

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