Myxofibrosarcoma directly invading superior vena cava causing obstruction successfully relieved by endovascular stent

JIHEON SONG[1], VINCENT M. MCDERMOTT[2], PAUL RYAN[3] & BRIAN R. HEALEY BIRD[1,4]

[1]*School of Medicine, University College Cork, Cork, Ireland,* [2]*Department of Diagnostic Imaging,* [3]*Department of Pathology, Bon Secours Hospital, Cork, Ireland and* [4]*Department of Haematology Oncology, Bon Secours Hospital, Cork, Ireland*

To the Editor,

Myxofibrosarcoma (MFS) is a soft tissue malignant neoplasm, seen predominantly in older people often occurring in the limbs but also rarely on the trunk, head and neck [1–3]. It has a high rate of local recurrence in spite of repeated surgical resection with wide negative margins [4]. However, MFS rarely metastasises and has not been reported to directly invade major vessels [5]. We describe an 80-year-old patient with MFS, where tumour directly invaded the superior vena cava (SVC). The diagnostic workup, palliation and pathogenesis of this tumour are discussed.

Case history

History, exam and diagnostic imaging

An 80-year-old female with a remote history of left-sided breast cancer was referred with a one-month history of indurated nodules on the right side of her neck. On examination, she had a palpable midline mass and associated right supraclavicular nodes. Recurrent breast cancer was suspected and an urgent ultrasound-guided core biopsy was performed, which yielded necrotic, non-diagnostic material. The patient was admitted 10 days later for further investigations. By this time, her face, neck and upper extremities (left more than right) were swollen and distended veins were newly visible over the chest and neck. Upon raising her arms, venous distension became more pronounced (Pemberton sign). She had deteriorated rapidly and was increasingly fatigued and weak.

CT scan (Figure 1) showed direct invasion and obstruction of SVC by tumour in lateral neck, possibly arising in a cervical lymph node, extending inferiorly into the superior mediastinum by intravascular spread, and superiorly obliterating the thyroid. Lung metastases and an intra-abdominal mass were noted.

Diagnostic procedure and pathologic findings

Another core biopsy was non-diagnostic and the Ear Nose and Throat service were reluctant to dissect the mass involving the carotid artery. As it was felt to be the most readily resectable lesion, without proximity to major blood vessels or vital structures, the patient underwent laparoscopic resection of a $7.2 \times 3.9 \times 3.5$ cm omental mass. Gross examination of the omental mass revealed a well-circumscribed, predominantly solid, focally cystic, cream-coloured variegated mass, focally haemorrhagic, with mucoid consistency and a maximum dimension of 4.5 cm (Figure 2). On microscopy the tumour was comprised of malignant spindle cells with myxoid differentiation, moderate nuclear pleomorphism, abundant mitoses, and foci of necrosis (Supplementary Figure 1a and b, available online at http://informahealthcare.com/doi/abs/10.3109/0284186X.2014.953261). On immunohistochemistry tumour cells were diffusely positive for vimentin and negative for epithelial (cytokeratins –

Correspondence: B. R. Healey Bird, Department of Haematology Oncology, Bon Secours Hospital, College Road, Cork, Ireland. Tel: + 353 21 4542807. Fax: + 353 21 4542350. E-mail: bbird@bonsecours.ie

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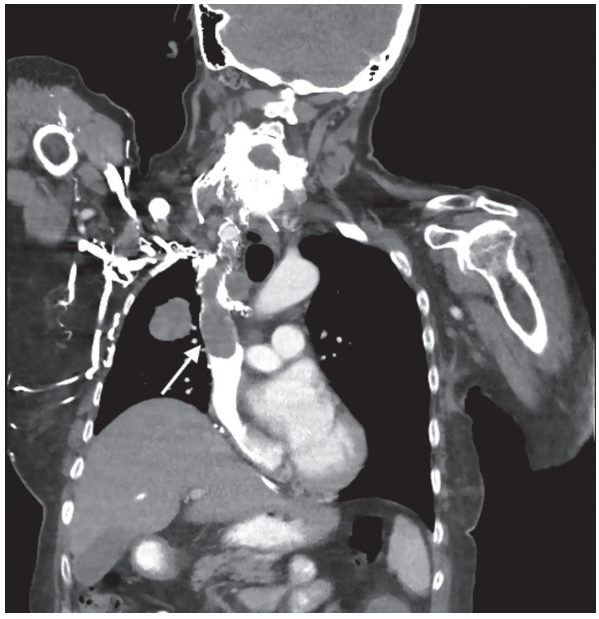

Figure 1. CT scan images of SVC obstruction (white arrow) by intravascular metastasis of MFS.

AE1/AE3, MNF116, CAM 5.2), melanocytic (S100, HMB45, Melan A), smooth muscle (caldesmon, desmin, actin) and various other markers including CD31, TTF-1, CD34, Bcl-2, and CD117. The morphological features and immunophenotype were compatible with high-grade sarcoma with myxoid differentiation, in keeping with a high-grade MFS. Repeat cytology from an ultrasound-guided percutaneous fine needle aspirate of the intravascular lesion revealed spindle cells similar to the omental mass.

Management of SVC obstruction

Upon CT scan results, steroids were started and therapeutic subcutaneous low-molecular heparin commenced. Once the chemotherapy insensitive nature of the tumour was confirmed, an SVC stent was placed under fluoroscopic guidance (Supplementary Figure 2a, b and Video 1–2, available online at http://informahealthcare.com/doi/abs/10.3109/0284186X.2014.953261), followed by immediate relief of vascular symptoms. Unfortunately, performance status failed to improve and she was transferred to in-patient hospice care three weeks post-admission and succumbed shortly thereafter.

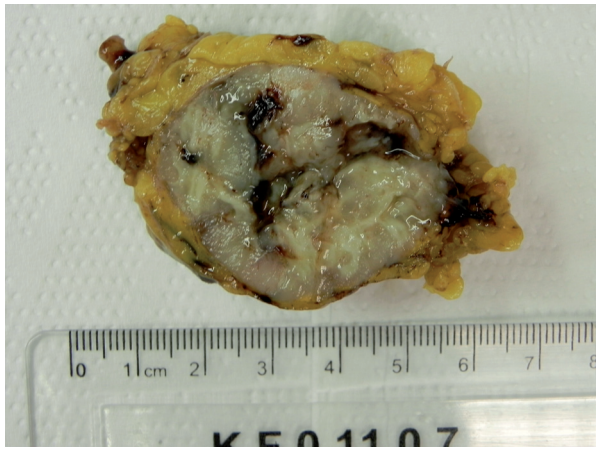

Figure 2. Gross appearance of omental mass: circumscribed, solid/cystic, variegated, focally haemorrhagic, focally necrotic (metastatic) tumour deposit.

Discussion

Previously classified as a myxoid variant of fibrous histocytoma, MFS has been recently reclassified as fibroblastic sarcoma due to its lack of histiocytic differentiation [6]. MFS usually presents as a lesion in dermal and subcutaneous tissues of the extremity and is treated by wide locoregional resection followed by adjuvant chemotherapy or radiotherapy [7,8]. However, the efficacy of adjuvant treatment is controversial [3,9]. Due to multiple metastatic sites at presentation, the primary site of our patient's MFS cannot be specified with certainty, but it is most likely to have arisen in the soft tissues of the right neck.

To our knowledge, this is the first report of MFS directly invading SVC. Sarcoma is a rare cause of SVC obstruction. In most cases, SVC syndrome (SVCS) results from compression of SVC by extravascular tissue mass, such as malignancy (65% of all SVCS cases are lung cancer or lymphoma) [10] and mediastinal lymphadenopathy. However, in rare cases, intravascular tumour invasion or venous thrombosis can cause SVCS [11]. SVCS from provoked thrombosis, caused by central venous catheters and pacemaker leads, results in a more acute course and requires immediate treatment. Common symptoms of SVCS are dyspnoea, acute chest pain, cyanosis, facial oedema, venous distension, light headedness and Pemberton sign [12]. Isolated case reports have detailed intravascular extension of sarcomas including pulmonary artery sarcoma [13] causing SVCS, but to our knowledge SVCS caused by MFS has not been reported.

Common causes of SVCS such as lymphoma or small cell lung cancer are usually chemosensitive and are rarely stented. In cases where steroids, chemotherapy or radiotherapy is unlikely to cause rapid tumour shrinkage, endovascular stent placement is the gold standard for the treatment of SVCS [14–16]. It is our practice to use low-dose, enteric aspirin for long-term thromboprophylaxis in patients with SVC stents. This is extrapolated from data in coronary artery stents and studies that have shown that aspirin alone may be effective in prophylactic measure [17,18]. In our patient, successful placement of endovascular stent provided immediate relief of

symptoms, but due to the extent of tumour spread and poor performance status the subsequent clinical emphasis was on palliation.

MFS, where resectable, should be removed with wide margins by experienced sarcoma surgeons in high-volume centres, followed by adjuvant radiotherapy. Better and less toxic systemic treatments are required for MFS, given the high rate of local recurrence in an elderly population. Pre-clinical research suggests that MET and other drugable oncogenes are activated in MFS [19], which offers hope that novel agents, such as crizotonib, may have a role to play even in non-ALK sarcoma [20]. Also, randomised controlled trials on the efficacy of aspirin compared to other antiplatelet drugs will be beneficial in the long-term care of cancer survivors with permanent endovascular stents.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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Supplementary material available online

Supplementary Figure 1–2 available online at http://informahealthcare.com/doi/abs/10.3109/0284186X.2014.953261.

Supplementary Video 1. Pre-operation. Video 2. Post-operation available online at http://informahealthcare.com/doi/abs/10.3109/0284186X.2014.953261.

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