Complete metabolic remission of an irresectable mediastinal solitary fibrous tumour with concurrent chemoradiation

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ABSTRACT

Solitary fibrous tumour is a rare mesenchymal tumour of uncertain origin that occurs most frequently in the pleura, although it has also been described in extraserosal sites. The biological behaviour of the tumour is unpredictable. The case history is described of a patient diagnosed with a large symptomatic irresectable mediastinal solitary fibrous tumour who achieved a clinical, radiological and metabolic response after concurrent chemotherapy and radiotherapy.

Solitary fibrous tumour is a rare mesenchymal tumour of uncertain origin that occurs most frequently in the pleura, although it has also been described in extraserosal sites. The biological behaviour of the tumour is unpredictable. Complete surgical removal of the tumour is the treatment of choice. There are currently few therapeutic options for patients presenting with irresectable tumours, and often only palliation is offered. We describe a patient who was diagnosed with a large symptomatic irresectable mediastinal solitary fibrous tumour who achieved a clinical, radiological and metabolic response after concurrent chemotherapy and radiotherapy.

CASE REPORT

A previously healthy non-smoking 58-year-old Caucasian man sought medical assistance for dyspnoea on exertion, cough, dysphagia and chest pain which was most noticeable in the supine position. For 6 months he had shown symptoms of cardiac arrhythmias and hypertension which were adequately treated with medication. There was no weight loss, nausea or fever. A chest radiograph showed a mass in the posterior mediastinum that was confirmed on the CT scan, and a positron emission tomography (PET) scan with ¹⁸F-fluorodeoxyglucose (FDG) showed a large mediastinal mass with intense slightly inhomogeneous FDG uptake, maximal standardised uptake value 20.2 (fig 1, upper row). Bronchoscopic examination was inconclusive. An attempt to obtain tissue by mediastinoscopy was not successful because of mediastinal congestion, so a CT-guided transthoracic needle biopsy was performed. Histological examination showed a tumour with a patternless growth architecture and branching haemangiopericytoma-like vessels (fig 2). The diagnosis of a solitary fibrous tumour (SFT) was made. The tumour was considered to be irresectable and a "wait and see" policy was adopted.

Five months later the patient presented again with increasing dyspnoea (grade 2 on the CTCAE 3.0 scoring system), grade 1 dysphagia, chest pain and cardiac arrhythmias and hypertension. His general condition remained good (WHO 1) and organ function was adequate. A new CT scan showed massive tumour progression with encasement of the mediastinal veins and trachea and dislocation of the oesophagus. After reviewing the literature, one case report was found of an abdominal SFT which responded to combined chemoradiation.¹ We therefore decided to treat the patient with concurrent chemoradiation: intravenous carboplatin AUC 5 mg/ml/min on day 1 plus intravenous etoposide 100 mg/m² on days 1, 2 and 3 with an interval of 3 weeks. Three cycles were delivered. Cisplatin was substituted by carboplatin because of a creatinine clearance of 67 ml/min. Concurrently with the first cycle of chemotherapy, radiotherapy was initiated to a total dose of 42 Gy in 28 twice-daily fractions of 1.5 Gy with an interval of 8 h between fractions. The radiation dose was limited to 42 Gy because a mean lung dose of 19.4 Gy had been reached. The patient tolerated the treatment very well with only grade 1 oesophagitis for less than 3 weeks. During chemoradiotherapy the dyspnoea and chest pain disappeared, and 3 months after completion of chemoradiation the patient had no symptoms. An FDG PET-CT scan showed no FDG uptake in the mediastinal tumour and marked volume reduction (fig 1, lower row), but it was still irresectable. There was diffuse mild FDG uptake in the right lung corresponding to an asymptomatic radiation pneumonitis. Six months after treatment there was still no change in response on the FDG PET-CT scan, and currently (8 months after treatment) the patient has no symptoms and is working full time (WHO 0).

DISCUSSION

SFTs were first described as a unique entity in 1932 by Klemperer and Rabin.² They are rare mesenchymal tumours of uncertain origin that are found mostly in the pleura but also in numerous extraserosal sites.³ SFTs are usually observed in middle-aged adults of 20–70 years (median 50 years) with no difference between the sexes. Most SFTs are slow-growing painless masses. Large tumours may give rise to compression symptoms or may be the source of paraneoplastic syndromes (hypertrophic osteoarthropathy, clubbing and/or hypoglycaemia due to the production of an insulin-like growth factor). They are usually found incidentally by chest radiography.

Figure 1 Upper row: ^{18}F -fluorodeoxyglucose (FDG) positron emission tomography (PET)-CT images before treatment showing a large mediastinal mass with intense slightly inhomogeneous FDG uptake and a paratracheal mass with intense FDG uptake. Lower row: FDG PET-CT images after concurrent chemoradiotherapy showing no FDG uptake in the mediastinal mass which appears completely normal. The diffuse mild FDG uptake in the right lung corresponds to radiation pneumonitis.

A striking feature of SFTs is their unpredictable biological behaviour. There is no strict correlation between histological morphology and behaviour. However, most of the histologically benign SFTs are non-recurring and non-metastasising lesions. Most histological malignant tumours (10–15%) behave aggressively and have a tendency to recur and to metastasise.⁴ In our case the tumour had no malignant features but the patient had progressive clinical symptoms.

All previous reports have agreed that the best predictor of a benign clinical course is the completeness of the tumour resection.^{3–5–8} However, in this case the tumour was irresectable. For cases in whom complete resection is not possible, information about treatment options and prognosis is sparse. Postoperative adjuvant therapy with radiotherapy, chemotherapy or both has been sporadically used in recurrent or systemic disease, but its benefit remains unclear. De Boer *et al* reported a

clinical and radiological response to neoadjuvant chemoradiation together with selective embolisation in a young man with a large irresectable SFT in the lower abdomen, after which the tumour could be removed.¹

Given this report and the need for treatment due to symptomatic tumour progression, we decided to give the patient concurrent chemoradiation. Three months after the end of treatment a clinical, radiological and metabolic response was observed on the FDG PET-CT scan. Six months after treatment the response remained unchanged and, at the time of submission of this report (8 months after the end of treatment), the patient has no symptoms and is working full time.

To our knowledge, this is the first report of an irresectable mediastinal SFT treated with concurrent chemoradiotherapy with a complete metabolic and clinical response. We conclude that, in patients with an irresectable (mediastinal) SFT, concurrent chemoradiation is a valuable alternative to a “wait and see” policy, certainly in patients with progressive symptoms.

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Figure 2 Histological examination of a needle biopsy specimen showing a solitary fibrous tumour with typically bland not atypical spindle cells, collagen deposition and haemangiopericytoma-like vessels. Stain: H&E.