CASE REPORT

Monsters do exist: an unusual case of chest pain

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SUMMARY

A 19-year-old man presented to the emergency department with a 3-week history of chest pain. Despite normal examination, bloods and observations of his chest X-ray revealed a large opacity in the middle and lower right zone. Further imaging revealed a multicystic mediastinal teratoma. He was admitted under the respiratory physicians who arranged further investigations and discussed his case with the lung and testicular multidisciplinary team. He was started on neoadjuvant chemotherapy with a plan for future definitive surgical resection. Four months later he suddenly deteriorated due to airway compression from the tumour. He underwent an emergency thoracotomy and tumour resection. He is currently doing well and is thought to have a good prognosis following complete resection of his tumour. This case report summarises this rare diagnosis and emphasises the need for careful evaluation of patients despite their initial well appearance, especially in patients who are reattending.

BACKGROUND

Mediastinal teratomas are uncommon and largely asymptomatic. Symptoms are variable and generally arise due to compression of adjacent structures. Diagnosis on clinical examination alone is not often possible, therefore, organising complementary investigations is important.¹

This is an interesting case highlighting the need to look outside the box once common causes are excluded, especially when patients are representing. The patient's initial well appearance with normal observations, blood tests and a history and examination suggestive of musculoskeletal pain could easily have led to a missed diagnosis had he not have had a chest X-ray. The duration of symptoms and lack of response to simple analgesia, despite the fact he appeared well, prompted this further investigation. I hope this case highlights this issue and prompts admitting doctors and general practitioners (GPs) to consider this when reviewing patients.

Most teratomas are benign but roughly 1–5% are malignant,² and malignant transformation is possible although very rare.³ It is therefore an important condition to be aware of, and referral to the appropriate teams for close clinical and radiological follow-up or definitive surgical excision is important.

CASE PRESENTATION

A 19-year-old student, originally from Kerala, south India presented to the emergency department reporting of a 3-week history of the right-sided anterior and posterior chest pain.

The pain was described as sharp in nature and was exacerbated by movement or palpation of the area. He admitted recently going back to the gym as he felt he was gaining weight. He denied any shortness of breath, cough or fevers. He had no risk factors for thromboembolism, no comorbidities and no significant family history. He had good functional capacity and enjoyed an active life.

Clinically, he appeared well. There were no significant findings on examination of his cardiovascular and abdominal system. His trachea was central and he had good bilateral air entry with equal chest expansion. Despite normal appearance to his chest, it was noted that he had palpable tenderness on the anterior aspect of his upper right chest. He was normotensive with oxygen saturations of 100% on air and a respiratory rate of 14. His admission ECG and bloods were normal.

Musculoskeletal chest pain was the initial working diagnosis but it was noted that 6 months earlier he had presented to his GP with a similar symptom and was prescribed ibuprofen, which eased his discomfort. He had had recurrent similar episodes and despite visiting his GP again, he no longer found the pain eased with simple analgesics. This factor prompted a decision for further investigations and a chest X-ray was requested.

INVESTIGATIONS

His chest X-ray found a large well-delineated opacity measuring at least 11.5 cm, occupying most of the right mid and lower zones (figure 1). There were no previous chest X-rays for comparison.

Owing to the findings on his chest X-ray, I re-examined the patient but found no abnormalities on examination of his thyroid, lymph glands and testicles.

Further imaging with a CT scan revealed a 9.9 cm×12.2×14.5 well-circumscribed mass in the right side of the anterior mediastinum (figure 2). The mass had a multicystic appearance with areas of coarse calcifications. The mass was expanding into the right hemithorax compressing the superior vena cava and right atrium. Compression atelectasis and a small pleural effusion were noted suggesting an element of tumour rupture. The findings were in keeping with a multicystic mediastinal teratoma.

As 5% of germ cell tumours are extragonadally located,¹ an ultrasound of his testicles was arranged. This found that both testes contained significant microcalcifications but no evidence of a testicular tumour.

Measuring the serum tumour markers (α -fetoprotein (AFP) and human β -choriogonadotropin (β -HCG) is important in the diagnosis and follow-up of mediastinal germ cell tumours. Patients with benign teratomas have normal

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Figure 1 The chest radiograph demonstrating a large well-delineated opacity measuring at least 11.5 cm, occupying most of the right mid and lower zones.

tumour markers unlike those with immature teratomas.⁴ His AFP was initially mildly elevated but subsequently returned to normal levels over the following 2 months. His β -HCG and lactate dehydrogenase were normal in keeping with a benign teratoma.

TREATMENT

The patient was discussed by the lung multidisciplinary team. He was diagnosed with a primary intrathoracic germ cell tumour (T4 N0 M0). He was referred to the cardiothoracic surgeons and oncologist and it was decided that he would start neoadjuvant chemotherapy prior to definitive surgical resection treatment.

His pain was managed with ibuprofen, paracetamol and codeine and he was discharged home with an outpatient follow-up in 1 week.

OUTCOME AND FOLLOW-UP

The patient received four cycles of BEP chemotherapy (bleomycin, etoposide, cisplatin). Despite chemotherapy, his repeat thorax CT revealed significant increase in the size of his tumour, which occupied virtually the whole right hemithorax and was causing significant mass effect on the mediastinum.

Figure 2 The CT scan that revealed a 9.9 cm×12.2 cm×14.5 cm well-circumscribed mass in the right side of the anterior mediastinum. The mass has a multicystic appearance with areas of coarse calcifications and is expanding into the right hemithorax compressing the superior vena cava and right atrium.

Four months later, while being treated in the hospital for an infection, he suddenly deteriorated and went into type 1 respiratory failure secondary to airway compromise from tumour compression. He underwent an emergency right thoracotomy and resection of his teratoma.

He is currently recovering on the intensive care ward, where he is thought to have a good prognosis as the surgical resection was complete.

DISCUSSION

The word 'teratoma' was first described in 1952 by Willis as 'a tumour composed of tissues foreign to the parts in which they arise'. It originates from the Greek word 'teras' which means monsters.⁵

It consists of multiple tissue types containing the three germ cell layers; endoderm, mesoderm and ectoderm. Ectoderm is the predominant tissue found; therefore, the finding of skin, hair and especially teeth is pathognomonic of the condition.

Mediastinal teratomas are rare accounting for only 5–10% of all mediastinal tumours.⁵ Most are benign, slow growing encapsulated tumours, with the majority being found incidentally in asymptomatic patients on X-ray.⁶

Symptoms arise as complications secondary to compression and erosion into adjacent structures. Chest pain is the most common symptom, but is atypical and can be sharp and constant. Rarely patients can present with coughing up of hair also known as trichoptysis due to erosion into the respiratory tract.⁵

Chest radiography is the initial step directing most patients for further investigation. CT is the investigation of choice as it determines the extent of the tumour and its relationship with adjacent structures. MRI and ultrasound are not used as primary diagnostic tools due to the detail provided by the CT scan.⁷

Tumour markers such as serum AFA and β -HCG levels should be measured as abnormal levels >500 mg/mL are diagnostic and suggest malignancy.⁷ Patients with preoperative normal tumour marker levels have been found to have better survival than those with elevated levels.⁸ Diagnosis is only confirmed by the presence of at least two of the germ cell layers taken from fine-needle aspiration or biopsy.

Surgical resection is the treatment of choice. Owing to the invading nature of teratomas, resection of associated intrathoracic structures may be necessary and must be considered when involving major structures such as the superior vena cava. Tumour recurrence following resection is rare.⁶

Neoadjuvant chemotherapy is effective and alongside rapid decreasing tumour marker levels are associated with a better overall survival.⁸

Learning points

- ▶ Mediastinal teratomas are rare but account for 5–10% of all mediastinal tumours.
- ▶ Clinical examination is usually normal and further imaging is required. CT of the thorax is the investigation of choice.
- ▶ Reattendance for the same problem, particularly when the initial treatment which had previously controlled the symptoms is no longer helping, should prompt a rethink and further investigations.
- ▶ As most are benign, encapsulated and slow-growing tumours, surgical resection is curative as recurrence is rare. Neoadjuvant chemotherapy has proven to be associated with a better overall survival.

Competing interests None.

Patient consent Obtained.

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