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Abstract

We describe the case of a 17-year-old hairdresser who presented with haemoptysis and trichoptysis due to benign intrapulmonary teratoma and her surgical management. The clinical and radiological features of this rare tumour are reviewed and the symptom of trichoptysis discussed.

Keywords: Intrapulmonary teratoma; Trichoptysis

1. Introduction

Teratomas are encapsulated germ cell tumours that contain tissue from one or more of the three germinal layers. They most commonly occur in the gonads but around 3% are known to be extra-gonadal [1], most of these being intrathoracic. We present a case of this very rare tumour with an extremely rare symptom, trichoptysis.

2. Case report

A 17-year-old hairdresser presented to an Emergency Department with a 2-day history of haemoptysis. She reported producing approximately half a cup full of blood with clots on several occasions. She also described a 6-month history of intermittent cough with expectoration of chalk-like material and hair. Interestingly, she had not reported this previously as she thought it was a normal occurrence for a hairdresser. She had otherwise been well and denied breathlessness, weight loss or pain.

At the time of admission, a chest radiograph identified a large left hilar mass which was confirmed on CT-scan (Fig. 1) and further characterized as being cystic and nodular with associated collapse and consolidation of the distal lung. A provisional diagnosis of teratoma was made, and serum germ cell tumour markers were tested and found to be negative. The patient then underwent a rigid bronchoscopy and left anterior mediastinotomy. Rigid bronchoscopy showed streaks of fresh blood originating from both upper lobe and lingular bronchi, which were cleared. Left anterior mediastinotomy revealed a large, encapsulated, predominantly left upper lobe mass which was biopsied and sent for histology. Importantly, the left anterior mediastinotomy enabled confirmation that the mass was primarily intrapulmonary with dense adhesions to the left phrenic nerve, making a left thoracotomy the surgical approach of choice should definitive resection be indicated. Macroscopically, the mass contained caseous material and hair while microscopically it exhibited evidence of a monodermal teratoma.

Ten days later the patient returned to theatre and underwent a left thoracotomy. The large mass was excised en bloc with the left upper lobe and phrenic nerve. The mass measured 70×50×60 mm with solid areas of sebaceous material and white hair as well as nodular, fatty protuberances (Fig. 2). There was no malignant or immature tissue identified and microscopic appearances were consistent with a mature teratoma.

The patient recovered well postoperatively and was discharged four days later.

3. Discussion

Intrathoracic teratomas are thought to be derived from the third pharyngeal pouch, a common origin they have with the thymus [2]. Evidence to support this theory has come from reports of thymic tissue found within resected teratoma specimens [3]. This suggestion has been used to explain why they seem to have a predilection for the anterior mediastinum [1]. In fact, germ cell tumours make up 15% of all anterior mediastinal masses that present in adults [4]. Mediastinal teratomas are subdivided into benign or malignant, with the malignant variety occurring almost exclusively in males [4].

Intrapulmonary teratomas (IPTs), in contrast, are significantly rare – 65 cases had been reported in total in Japanese and English literature up to 1996 [2]. IPT typically present between the first and second decades of life, and unlike malignant mediastinal germ cell tumours that have male predominance, IPTs have an equal distribution amongst males and females [2]. Previous studies have found that IPTs tend to occupy the left upper lobe of the lung...
Fig. 1. Preoperative CT-scan demonstrating the heterogenous appearance of the teratoma.

Fig. 2. Resected teratoma specimen showing the white hair within the tumour mass.

[5], as demonstrated in our case, the reasons for which a widely accepted explanation is yet to be provided. The presenting symptoms for IPTs are variable. Most common are chest pain, haemoptysis and cough but these are all common symptoms making preoperative diagnosis difficult. However, most literature sites the pathognomonic symptom of IPT as expectoration of hair, or trichoptysis. This highly specific symptom occurs in 13% of IPTs and is due to a direct communication between the teratoma and the bronchus [6]. Interestingly, it has been well documented, and noted in our case, that the expectorated hair in this condition is commonly white in appearance [7]. This may be a reflection of the lack of maturity of the pigment producing cells within the tumour. A review of the literature reveals only one other reported cause for trichoptysis not related to IPT but instead acquired as a complication of reconstructive surgery for a laryngeal tumour [8]. Although hairdressers do report increased upper and lower respiratory tract symptoms [9], trichoptysis should not be one of them!

A well-documented risk associated with this tumour is that of rupture which is proposed to be due to proteolytic or digestive enzymes derived from the tumour [10]. Rupture into the pericardium, mediastinum or bronchial tree may produce cardiac tamponade, granulomatous mediastinitis or lipid pneumonia, respectively [2].

The wide variety of symptoms by which this condition presents make the clinical diagnosis of IPT extremely difficult and hence heavily reliant on radiological investigations. The evidence provided by CT-scanning is invaluable and serves two important purposes. Firstly, CT assists in making the diagnosis of this rare tumour with, as already mentioned, clinically indistinguishable characteristics. Secondly, CT helps determine whether the tumour has ruptured or not [1].

Although the risk of rupture has been agreed to be rare [10], the significance of preoperative diagnosis is underlined by the associated inflammation and adhesion caused by leakage of internal components of a ruptured teratoma that make surgical intervention more complex [10]. As with most tumour markers, beta HCG and AFP are limited by their low specificity, and positive results merely complement clinical and radiological findings.

Surgery is the mainstay of treatment for IPT. For unruptured teratomas, this is done mainly to relieve symptoms, particularly haemoptysis, and to curb the potential risk of malignant change of the tumour [5]. Surgery of the ruptured teratoma aims to deal with the complications already described and avoid the ultimate complication of adult respiratory distress syndrome [10].

4. Conclusion

In conclusion, IPTs are rare germ cell tumours whose symptoms at presentation are mostly non-specific apart from that of trichoptysis, which although rare, is pathognomonic. The diagnosis of IPT is dependent on careful clinical consideration and the use of CT imaging, with surgical resection being the mainstay of its treatment.

References

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