Photoclinic



Figures 1. Presence of hair, sebum, and teeth as seen in expectorted material.

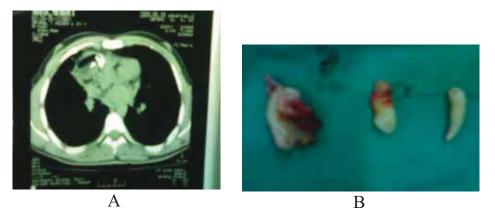


Figure 2. A) CT scan shows a mediastinal mass and teeth, B) three teeth postoperatively.

A 22-year-old man referred to the Pulmonary Department, Razi Hospital, Rasht in Guilan Province with a two-week history of hemoptysis. He had history of around 20 mL of bloody sputum with clots on several occasions. The patient also described a six month history of intermittent cough with expectoration of hair and sebum (Figure 1A). Prior to his visit, he reported expectoration of teeth-like material (Figure 1B). Otherwise he was well and denied symptoms of breathlessness, weight loss or pain. Physical examination was normal. All laboratory tests also were within normal limits. Chest radiograph showed a large mediastinal mass which involved the right upper lobe of lung and

Manoucheher Aghajanzadeh MD¹, Ali Alavi MD¹, Gilda Aghajanzadeh MD¹, Sara Massahnia BS

•Corresponding author and reprints: Manoucheher Aghajanzadeh MD, Respiratory Diseases & TB Research Center, Razi Hospital, Guilan University of Medical Sciences (GUMS), Rasht, Iran. P.O. Box: 41448 Tel: +98-113-155-42460; Fax: +98-113-155-59787; E-mail: smassahnia@yahoo.com was confirmed on CT scan (Figure 2A). The mass was characterized with cavitation and two bony-like appearances, with associated collapse and consolidation of the distal lung. Serum germ cell tumor markers were tested and found to be negative. The patient underwent fiberoptic bronchoscopy which showed streaks of fresh blood originating from right upper lobe. Right anteriolateral thoracotomy revealed a large mass which predominantly had involved the right upper lobe. The large mass was excised en bloc. Macroscopically, the mass contained hair with three teeth (Figure 4). Microscopically, there was no sign of malignant or immature tissue to be identified. The patient recovered well postoperatively and was discharged six days later.

> What is your diagnosis? See the next page for diagnosis.

Authors' affiliation: 'Respiratory Diseases & TB Research Center, Razi Hospital, Guilan University of Medical Sciences, Guilan, Iran.

Accepted for publication: 2 June 2010

Teratoma

Photoclinic Diagnosis:

B enign germ cell tumors are uncommon and account for 5 to 10% of all mediastinal tumors¹ Intrathoracic teratomas are thought to be derived from the third pharyngeal pouch, a common origin with the thymus.² Evidence to support this theory has come from reports of thymic tissue found within resected teratoma specimens.³ Only 5 to 10% of germ cell tumors are extragonadal, however the mediastinum is the most common extragonadal site. Typically, germ cell tumors occur in young adults in the second to fourth decades of life with equal sex distribution,^{1,4} which was the case with the patient.

Histologically benign teratomas consist of all three primordial layers: ectoderm (skin and hair), mesoderm (bone, fat and muscle) and endoderm (respiratory epithelium and gastrointestinal tract). Skin, pilosebaceous tissue, smooth muscle, fat and respiratory epithelium occur in more than half of the tumors.⁵⁻⁶ Teeth, bone, cartilage, neural tissue and pancreatic tissue have been reported in benign tumors.¹ In this case, all three layers were present. The tumor tends to be slow growing, thus many patients have no signs or symptoms when the mass is initially discovered.^{1,4} The most common symptom is chest pain, which may be accompanied by dyspnea, cough, and fever.1 Chest pain is most commonly substernal or in the pectoral area and located on the side where the mass is present. Occasionally, when the tumor erodes into an airway patients may cough up hair (trichophytosis) or sebum, which is pathognomonic of a benign mediastinal teratoma.⁵ Hemoptysis can occur, presumably from irritation of the bronchial mucosa by the tumor or from erosion into a vascular structure. Presenting symptoms also can occur from pressure on surrounding structures.^{1,5} The patient had rare presentations of cough, hemoptysis, expectoration of hair and sebum, and teeth-like material.

Benign teratomas, in addition to eroding into an airway, can also rupture and fistulize into the pleural or pericardial spaces and present with cardiac tamponade.^{1,5} Benign teratomas rupture into the lung or bronchia tree and produce hemoptysis.¹

Obviously, symptoms depend on what structure the tumor ruptures into. If the tumor ruptures in the lung, an abscess or coughing of hair or sebaceous material can result. If the teratoma erodes into the pleural space, a pleural empyema can develop which present with chest or back pain.¹ Most benign teratomas are discovered on routine chest radiography. CT scan with contrast enhancement is the diagnostic procedure of choice to evaluate mediastinal abnormalities found on chest radiography.^{1,6,7} Usually, a CT demonstrates a thick-walled cystic mass. When areas of calcification are intermixed within areas of fat density, the diagnosis of teratoma can be made with a high degree of certainty.¹ The role of magnetic resonance (MRI) imaging in evaluating teratomas is not fully known, but the ability to determine vascular structures without contrast may be a distinct advantage.¹ Tumor markers should be obtained in all young adult patients diagnosed with an anterior mediastinal mass.¹

The most common markers that should be obtained include n-fetoprotein and 13-human chorionic gonadotropin.1 The treatment of choice for benign germ cell tumors is total excision before complications ensue. Resection may be accomplished via a median sternotomy, lateral thoracotomy, or, rarely, a clamshell incision.^{1,8} The patient underwent a right anteriolateral thoracotomy for mediastinal mass and right upper lobe resection. If necessary, complete excisionof thymus, lung and pericardium should be performed, but tissue may need to be left behind to avoid injury to vital neural or vascular structures.⁹ If the tumor has fistulized to the lung, a pulmonary resection may be necessary.¹ The prognosis after excision is excellent even when complete excision is impossible.5 Postoperative irradiation or other adjuvant measures are not indicated in any of these patients.¹ The ten year survival rate is 92.8%.^{1,6}

References

- Allen MS, Trastek VF, Pairolero PC. Benign germ cell tumors of the mediastinum. Editors: Shields TW, LoCicero J, Reed CE, Feins RH, eds. *General Thoracic Surgery*. Vol. 3, 7th ed. London; Lippincott Williams; 2009: 2389.
- Rana SS, Swami N, Mehta S, Singh J, Biswal S. Intrapulmonary teratoma: an exceptional disease. *Ann Thorac Surg.* 2007; 83: 1194–1196.
- Duwe BV, Sterman DH, Musani AI. Tumours of the mediastinum. Chest. 2005; 128: 2893 – 2909.
- Agarawal R, Srinivas R, Saxena AK. Trichoptysis due to an intrapulmonary teratoma. *Resp Care*. 2007; 52: 1779 – 1781.
- Chen RF, Chang TH, Chang CC, Lee CN. Mediastinal teratoma with pulmonary involvement presenting with haemoptysis. *Chest.* 2007; 132: 683 – 685.
- Tafadzwa P, Makarawo Finnikin S, Woolley S, Bishay E. Trichoptysis: a hairy presentation of a rare tumor. *Interact CardioVasc Thorac Surg*. 2009; 9: 733 – 735.
- Bakry M, Arshad S, Haq S, Gebreab F, Voigt L, Raoof S, et al. Hairy hoarseness. *Chest.* 2004; **126**: 943 – 947.
- Choi SJ, Lee JS, Song KS, Lim TH. Mediastinal teratoma: CT differentiation of ruptured and unruptured tumours. *Am J Roentgenol.* 1998; 171: 591 – 594.
- Zenker D, Aleksic I. Intrapulmonary cystic benign teratoma: a case report and review of the literature. *Ann Thorac Cardiovasc Surg.* 2004; 10: 290 – 292.