CASE REPORT

Rhabdomyoma as a tumor of the posterior mediastinum

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KEY WORDS

mediastinum, rhabdomyoma, tumor

ABSTRACT

Rhabdomyoma is a rare tumor in the posterior mediastinum. We present a case of adult rhabdomyoma that occurred multifocally in the head, neck and mediastinum. There have been as few as four previous reports on rhabdomyoma in the posterior mediastinum. An 80-year-old white man was admitted to the Division of Pulmonary Diseases because of dyspnea, hemoptysis and fatigue. Physical examination revealed a tumor in the left submandibular area. Computed tomography (CT) of the head and neck showed a mass which caused a marked prominence of the left posterolateral wall of the pharynx. Chest CT revealed a contrast-enhancing tumor. The mass was 20 × 19 × 55 mm in size and was in contact with the lateral wall of the esophagus. Histological examination showed that the tumor was composed of cells typical of adult rhabdomyoma. The patient has been followed in our outpatient clinic for more than the last 7 years. To our knowledge, this is the fifth case of adult mediastinal rhabdomyoma. We believe that rhabdomyoma is a rare tumor which should be considered in a differential diagnosis of tumors detected in the posterior mediastinum.

INTRODUCTION Tumors or cysts located in the mediastinum are lesions which originate from its structures. Metastases or tumors involving the mediastinum but originating from other intrathoracic sites are not classified as mediastinal tumors. Similarly, primary tumors of the trachea, esophagus and heart are not included. Tumors of the posterior mediastinum are most frequently neurogenic tumors arising from peripheral nerves and sympathetic ganglia.[1,2]

Rhabdomyoma is a rare benign neoplasm of soft tissue. It accounts only for 2% of all tumors with skeletal muscle differentiation.[3] These tumors may originate from cardiac or skeletal muscles. Rhabdomyomas may be cardiac or extracardiac. Cardiac rhabdomyomas are the most common primary tumors occurring in newborns and children. They are often associated with tuberous sclerosis and are considered hamartomatous. Extracardiac rhabdomyomas are classified into three clinical and histological subtypes: adult, fetal and genital. They account for 50%, 40% and 10% of extracardiac rhabdomyomas, respectively. Extracardiac adult rhabdomyomas (EAR) are located in 70–75% of cases in the head and neck region. The most common site is the mucosa of the pharynx, oral cavity and larynx. These tumors have been rarely reported to occur in the extremities, esophagus, stomach, and mediastinum. The fetal subtype may occur in children and adults, typically in the head and neck region. The genital subtype is a polipoid tumor which occurs in the vagina or vulva in middle-aged women.

To our knowledge, our patient is the fifth case of adult rhabdomyoma occurring in the mediastinum[3-6] and the first case of multifocal rhabdomyoma detected in the head, neck and mediastinum.

CASE REPORT An 80-year-old white man was admitted to the Division of Pulmonary Diseases, Kraków, Poland. He complained of dyspnea, weakness, sweating (mainly in the neck region), dry and persistent cough, sore throat, and subfebrile state. Hemoptysis and epistaxis occurred 2 days before admission.

The patient suffered from chronic obstructive pulmonary disease and ischemic heart disease. At the age of 33 he underwent tonsillectomy. At the age of 50, 53 and 63 he expierenced spontaneous pneumothoraxes treated with tube drainage. At the age of 63, he underwent

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cholecystectomy due to symptomatic cholelithiasis. Frequent upper respiratory tract infections have been observed over several years.

On admission, a physical examination revealed a large tumor in the left submandibular area. Laboratory tests showed only elevated sedimentation rate (40/h).

Ultrasound examination of the neck showed hypoechogenic mass, with echogenic septa, extending laterally (50 × 40 mm). A few hypoechogenic, well-vascularized, oval lesions were also detected in the submental region, the largest 14 × 13 × 22 mm in size. They were similar to lymph nodes in terms of appearance and perfusion. Computed tomography (CT) of the head and neck confirmed the presence of a large tumor, which caused a marked prominence of the left posterolateral wall of the pharynx. The tumor was extending from the soft palate toward the epiglottis. It gradually moved laterally, enveloped the hyoid bone and was localized subcutaneously at the level of the epiglottis. The contrast-enhanced mass (up to 120 Hounsfield units [HU]) was homogeneous. It was well circumscribed and did not invade other organs. CT of the neck confirmed the presence of three other oval, contrast-enhancing lesions (up to 120 HU), with a maximum diameter of 16 mm (FIGURE 1A).

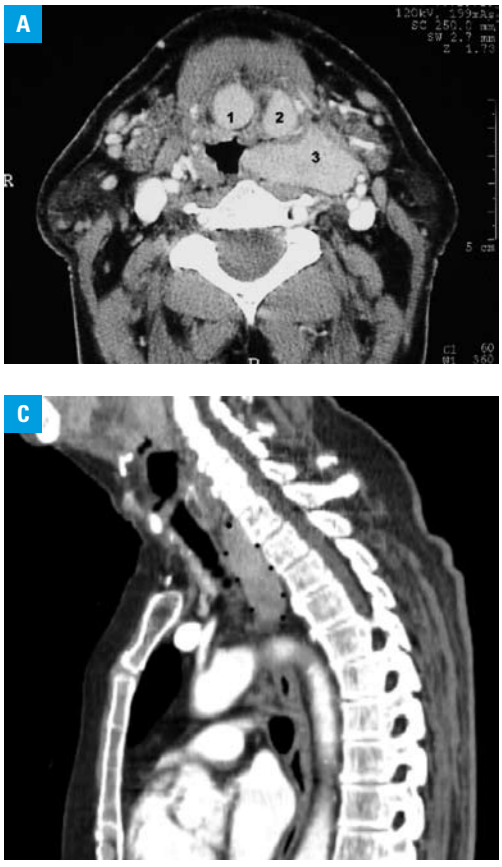
Chest radiograph showed no abnormalities. Chest CT with intravenous contrast medium revealed a contrast-enhancing tumor (up to 120 HU) in the posterior mediastinum. The tumor size was 20 × 19 × 55 mm and it was in contact with the left lateral wall of the esophagus (FIGURE 1B, C, D). It had regular margins and was homogenous. There was no mediastinal lymphadenopathy.

Bronchofiberoscopy showed no significant abnormalities. Mucosa of the medial wall of the right main bronchus was slightly damaged and covered with fibrin. Hemoptysis might have resulted from bleeding from this site.

The patient was referred to a laryngology department for further diagnostic procedures. Surgical biopsy of the neck tumor was performed. Cells of adult rhabdomyoma were found on histological examination. The patient was referred to a thoracic surgeon. However, a surgery was not performed due to a benign and multifocal nature of the neoplasm. He did not give consent to undergo gastroscopy.

The patient has been followed in our outpatient clinic for the last 7 years. He complains of recurrent productive cough with purulent sputum, dyspnea on exertion, the sensation of hindrance in the throat, mild dysphagia, and hoarseness. The tumors are stable on a repeat CT scan of the neck and chest.

DISCUSSION EARs are extremely rare neoplasms, affecting patients aged 40–70 years.[3] They are observed more often in men than in women,with the male to female ratio of about 3–6 to 1. They are typically found in the region of the head and neck. An EAR is usually unifocal, but 15% of the cases are multifocal.[5] These tumors usually



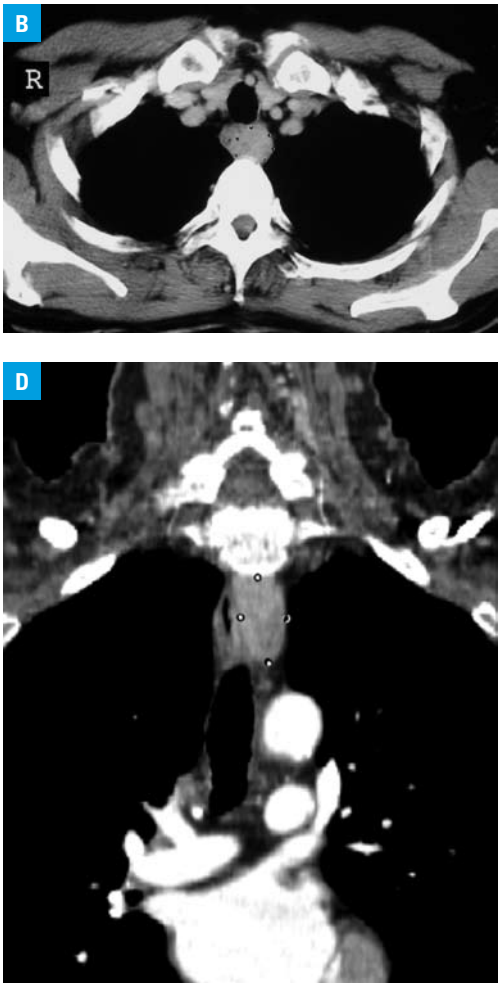


FIGURE 1 Radiological images of a rhabdomyoma
A computed tomography (CT) of the head and neck confirmed the presence of a large tumor (52 × 24 × 58 mm) (3) and other oval lesions (1,2)
B chest CT – axial scan, a tumor (20 × 19 × 55 mm) in the posterior mediastinum
C chest CT – sagittal reconstruction, a tumor in the posterior mediastinum
D chest CT – frontal reconstruction, a tumor in the posterior mediastinum causing compression and a shift of the esophagus to the right

cause symptoms by compressing the neighboring organs. Symptoms depend on the size and location of the lesion and typically include airway obstruction, dysphagia, hoarseness, and nasal obstruction.

So far, four cases of an EAR located in the mediastinum have been reported. The first case was described in 1978.[6] An 80-year-old male presented with swallowing difficulties. An EAR was diagnosed at autopsy. The tumor was located in the anterior-superior mediastinum extending to the neck. Its size was 100 × 50 × 30 mm. In 1995, Box et al.[4] described a 52-year-old man with a recurrent respiratory airway infection and cough. A tumor in the posterior mediastinum was found on chest radiography. Chest CT showed that it extended to the neck and partially enveloped the trachea and esophagus. The patient underwent a surgery. Due to the size of the tumor thoracotomy and cervical incision were performed. The patient recovered and no symptoms were observed after 8 months. In 2002, an anterior-superior mediastinal rhabdomyoma was reported[5] in a 68-year-old man who had been killed by a moving motor vehicle. The tumor was detected at autopsy and there were no data on the patient's symptoms. Similarly to the previous cases, the tumor was lobulated. Its size was 63 × 45 × 30 mm and was surrounded by a thin capsule. In 2006, the fourth case of EAR in the mediastinum was reported.[3] A 75-year-old woman was diagnosed with recurrent fever. There was a tumor in the posterior mediastinum behind the trachea and esophagus, extending to the prevertebral space of the neck. A cervical incision and partial upper sternotomy were performed. The tumor was completely removed. It was lobulated, had a size of 160 × 87 × 25 mm and weighed 100 g. It was surrounded by a thin capsule. The patient made full recovery. A 10-month follow-up was uneventful.

Unlike the previously reported cases, an EAR in our patient is multifocal. It is also the first mediastinal EAR, which has been observed for several years. We are unable to determine when these tumors developed in the patient but we have observed no enlargement for 7 years.

An EAR is a benign neoplasm, which consists of cells with eosinophilic granular cytoplasm resembling cross-striations of myofibrils. EAR cells contain desmin, muscle specific actin and myoglobin. Immunocytochemistry showed fetal myosin.

The diagnosis is established by fine-needle biopsy or surgical biopsy and subsequent histological examination of the tumor. In our case the lesions were extensive and multifocal. The diagnosis was based on histopathological examination of a single biopsy specimen and CT findings. Despite their varying sizes, the tumors did not show features of invasion. The margins were well-defined and regular. The density of changes, particularly the homogenous and intensive contrast enhancement (up to 120 HU) were indicative of a similar structure of all described changes. The stable tumor size during the 7-year follow-up is an essential marker of a benign nature of the tumors.

A complete resection is the treatment of choice for an EAR.[7] Tumor recurrence are usually associated with incomplete resection, therefore it is vital to completely remove the lesion.[7] Tumors grow slowly and their clinical manifestations are late. Patients after resection require regular follow-up visits.

Rhabdomyoma, although detected extremely rarely, should always be considered in a differential diagnosis of tumors observed in the posterior mediastinum.

REFERENCES

1 Sladek K. [Mediastinum diseases]. In: Szczeklik A, ed. [Internal Medicine]. Kraków, Medycyna Praktyczna, 2005: 643-644. Polish.

2 Park D, Vallieres E. Tumors and cysts of the mediastinum. In: Mason RJ, Broaddus VC, Murray CF, Nadel JA, eds. Textbook of Respiratory Medicine. Philadelphia, Elsevier, 2005: 2001-2038.

3 Zolota V, Tzelepi V, Charoulis N, et al. Mediastinal rhabdomyoma: case report and review of the literature. Virchows Arch. 2006; 449: 124-128.

4 Box JC, Newman CL, Anastasiades KD, et al. Adult rhabdomyoma: Presentation as a cervicomediastinal mass (case report and review of the literature). Am Surg. 1995; 61: 271-276.

5 Sidhu JS, Nicolas MM, Taylor W. Mediastinal rhabdomyoma: a and review of the literature. Int J Surg Pathol. 2002; 10: 313-318.

6 Miller R, Kurtz SM, Powers JM. Mediastinal rhabdomyoma. Cancer. 1978; 42: 1983-1988.

7 Cieslik T, Jędrzejewski P. [Rhabdomyoma – rare tumor of head and neck area – literature review]. Dent Med Prob. 2004; 41: 91-94. Polish.

OPIS PRZYPADKU

Mięśniak prążkowanokomórkowy jako przyczyna guza śródpiersia tylnego

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guz, mięśniak prążkowano-komórkowy, śródpiersie

STRESZCZENIE

Mięśniak prążkowanokomórkowy jest rzadką przyczyną guzów śródpiersia tylnego. Przedstawiamy przypadek mięśniaka prążkowanokomórkowego typu dorosłych występującego wieloogniskowo w obrębie głowy, szyi i śródpiersia. Do tej pory opisano tylko czterech chorych z mięśniakiem prążkowanokomórkowym śródpiersia tylnego. 80-letni chory został przyjęty do Kliniki Pulmonologii z powodu duszności, krwioplucia i osłabienia. W badaniu przedmiotowym uwagę zwracał guz okolicy podżuchwowej lewej. W tomografii komputerowej głowy i szyi stwierdzono zmianę uwypuklającą tylno-lewoboczną ścianę gardła. Tomografia komputerowa klatki piersiowej uwidoczniła w śródpiersiu tylnym heterodensyjny obszar ulegający wzmocnieniu po podaniu środka cieniującego. Zmiana o wymiarach 20 × 19 × 55 mm przylegała do bocznej ściany przełyku. W badaniu histopatologicznym zmiany stwierdzono utkanie typowe dla mięśniaka prążkowanokomórkowego typu dorosłych. Od ponad 7 lat chory pozostaje w obserwacji. Według naszej wiedzy jest to piąty opisany na świecie przypadek mięśniaka prążkowanokomórkowego występującego w śródpiersiu. Uważamy, że w diagnostyce różnicowej guzów śródpiersia tylnego należy uwzględnić rzadko występującą postać mięśniaka prążkowanokomórkowego.

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