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Centrally-located pulmonary hamartoma diagnosed in a 16-year-old boy presenting with chronic chest pain: a case report

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Abstract
Pulmonary hamartomas are the most common benign tumors of the lung. They are usually diagnosed incidentally while evaluating for other conditions. These tumors have been shown to be uncommon below the age of 25 years. We report a case of a 16-year-old male who presented with chronic chest pain and was histologically confirmed to have pulmonary hamartoma. The tumor was successfully resected. This is the first case of hamartoma to be reported in our country, and the atypical age at presentation together with the tumor's position makes it more unique. In conclusion, although very rare, pulmonary hamartomas can occur in young age with the central location. Clinically, the presentation is usually non-specific and the diagnosis is mostly incidental while investigating other conditions. There is a need to increase awareness among clinicians on hamartomas to improve early treatment for this rare disease in the adolescents.

Keywords: pulmonary, hamartoma, chest pain, Tanzania

Introduction

Hamartomas, also known as mesenchymomas, are non-neoplastic tumor-like malformations of the lung (Bateson, 1973). Up to 80% of these tumors can present as parenchymal lesions and only 20% may present as endobronchial (Cosio *et al*., 2002). In most cases, parenchymal lesions are found incidentally while taking chest radiographs for other reasons as rounded homogenous opacities at the periphery (Salminen, 1990). On the other hand, endobronchial lesions may present with new onset of respiratory symptoms including recurrent respiratory tract infections, obstructive pneumonia or haemoptysis (Cosio *et al*., 2002).

The peak incidence of these lesions is between the sixth and seventh decade of life with male preponderance (Gjevre *et al*., 1996). Occurrence in childhood is rare. Hamartomas have been found to be the most common benign tumors of the lung and account for up to 3% of all tumors of the lung (Schwartz, 1961) and up to 6% of all cases of solitary pulmonary nodule (Bateson, 1965). Histologically, hamartomas are usually composed of a mixture of cartilage, fat, fibrous tissue, smooth muscle and branching clefts of non-neoplastic reactive type epithelium. They can also be composed of adipose tissue with leiyomyomatous differentiation (Ganti *et al.,* 2006).

Despite of the advancement in medical therapy, pulmonary resection remains the mainstay for the treatment of the lung hamartoma. However, it is still unclear on when should the surgery be performed (de Rooij *et al*., 1988; Salminen, 1990). The issue of malignant change and recurrence

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postoperatively is also not clearly defined (Basile, 1989). Recurrence of the lesions has been shown to be common post surgical enucleation (Cosío *et al*., 2002).

Radiographic features

Pulmonary hamartoma are typically well-circumscribed nodules with either smooth or lobulated margins. Approximately 60% have fat (Klein & Braff, 2008) and approximately 20-30% have calcification (usually pop-corn like) (Chai & Patz, 1994). Cavitation is extremely rare. Size is variable, and they can be large (>10cm) but in most cases they are less than 2.5 - 4 cm in diameter. Chest radiography is non-specific, demonstrating soft tissue attenuation, well-circumscribed mass with either smooth or lobulated margins. Calcification (classically popcorn type) may be seen, which can suggest the diagnosis. Fat is difficult to identify with certainty on chest radiography. Computed tomography (CT) scanning is more sensitive than chest radiography in detecting fat and calcification. The reported prevalence of calcification in hamartomas on CT varies from 5 to 50% while fat may be identified in up to 50% of hamartomas on CT (Gaerte *et al*., 2002; Yilmaz *et al.*,2004; Karabulut *et al.*, 2007;) Hamartoma may be confidently diagnosed when a sharply marginated, smooth lesion containing calcification and fat, which does not demonstrate significant growth, is identified on a CT scan (Klein & Blaff, 2008).

Case presentation

We are reporting a case of 16 years old male patient, seen in a Medical outpatient department, at Bugando Medical centre, Mwanza-Tanzania with complaints of long-standing chest pain. The chest pain was reported to be on and off for approximately one year prior to seeking medical attention. It was of insidious onset, slowly progressive, and more intense on the left side of the chest. The chest pain was reported to worsen on exertion and was non-radiating. It was neither postural nor pleuritic. There were no fevers, cough or awareness of heart beat reported.

Review of other systems was uneventful. The patient had no significant past medical history with no known food or drug allergies. He was a first born in family of three, none with similar complaints. He lived with parents and he was a secondary school student at the time of presentation. He denied smoking cigarettes.

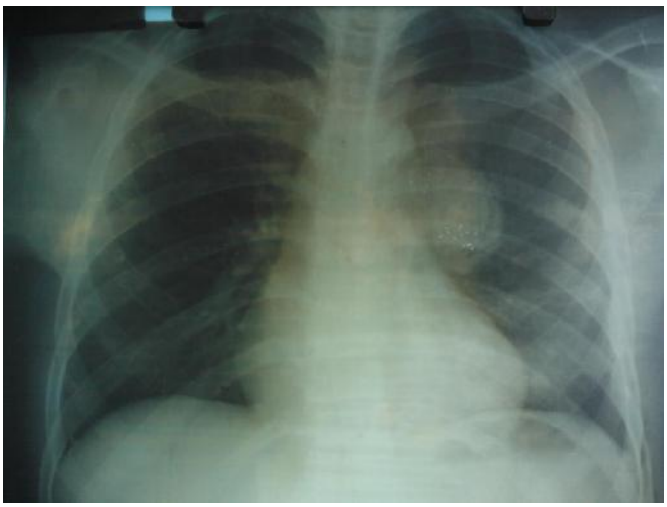

Figure 1: A Postero-anterior chest X-ray

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On general examination, the patient was alert, afebrile, not pale, not dyspnoeic with SPO_2 of 97% in the room air. The lymph nodes were not palpable and he did not have lower limb edema. Vital signs included a blood pressure of 120/80mmHg, heart rate 76 beats/minute, respiratory rate of 16 cycles per minute and the body temperature of 37°C. Nothing significant was found during the systemic examination. The patient was evaluated for a possibility of having mitral valve prolapse (MVP), and a chest radiography and echocardiogram were then ordered. The echocardiogram confirmed the presence of MVP. However, the chest film revealed a well circumscribed, rounded opacity on the left peri-hilar region, with characteristic popcorn calcification (Figure 1).

A chest computed tomography (CT) scan was then done which revealed rounded, smooth and sharply emarginated lesion on the left peri-hilar region, measuring 3cm, with multiple foci of fat and areas of calcification (Figure 2).

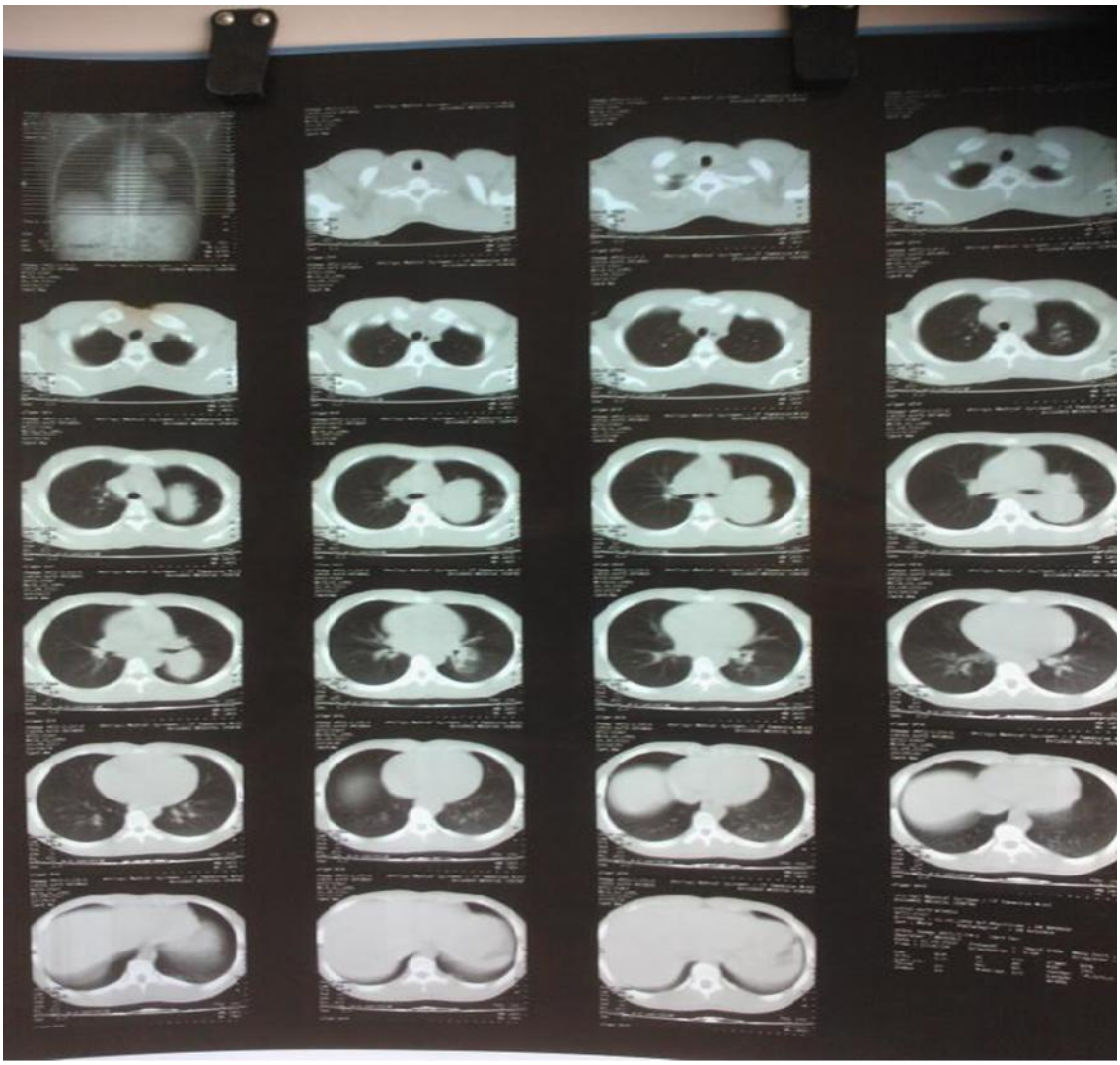

Figure 2: Non -enhanced, axial computed tomography scan of the chest

Thoracotomy was done following the cardiothoracic surgeon assessment. A mass measuring approximately 34mm by 32mm (approximately half the diameter of a tennis ball) was found and was fully resected. This mass was histologically confirmed to be a hamartoma. The patient was discharged

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one week post-surgery in good condition. Follow up was done after two weeks, and monthly for three months and no new developments were found. The patient was seen again two years post-operatively, at which time the control chest X-ray and the CT scan were all normal and he was asymptomatic.

Discussion

Pulmonary hamartomas are the most common benign tumors of the lung with a documented incidence of 0.025-0.32% (Murray *et al.*, 1991). They have been shown to contribute up to 3% of all lung tumors (Schwartz, 1961) and up to 6% of all solitary pulmonary nodules (Bateson, 1965). In literature, there are very few reports, most of which are old, and to the best of our knowledge there is no report published from Tanzania on this condition.

These tumors have been shown to predominantly affect individuals between the 5th and 7th decades of life with a male preponderance documented to range from 3-5:1 (Cosio *et al.*, 2002; Guo *et al.*, 2008). The tumor has been associated with smoking, with one report showing that more than 80% of the patients were smokers (Gjevre *et al.*, 1996). Occurrence has been shown to be very rare before the age of 30 years (Guo *et al.*, 2008). Our patient was 16 years at the time of diagnosis.

Pulmonary hamartomas can occur anywhere in the lung field, but the commonest site has been shown to be at the periphery. A hilar location, as was the case with our patient, is much rarer (Bateson, 1965). In the vast majority of cases, pulmonary hamartomas are solitary (Bini, 2002; Kang, 2007). These tumors can either be parenchymal or endobronchial in origin with parenchymal being the commonest type. Only a few cases of pulmonary hamartomas have been shown to be endobronchial (Leroux, 1964; Sibala, 1972; Gjevre *et al*. 1996).

The clinical manifestation of pulmonary hamartomas has been shown to be non-specific with most patients being diagnosed coincidentally on chest radiographs taken to evaluate other conditions (van den Bosch, 1987; Salminen, 1990; Gjevre *et al*., 1996; Guo *et al.*, 2008). Our patient presented with chest pain, which could have been due to the MVP that was found on echocardiogram. However, his pain could have been caused by the hamartoma as it disappeared immediately post surgical resection. The MVP was not corrected in this patient.

Surgical resection remains the mainstay of treatment for pulmonary hamartomas. Among the indications for surgical resection is a tumor diameter of more than 2.5cm (Guo, 2008). Our patient had a tumor with a diameter of 3.5cm. It has been reported that recurrence of tumors post surgical resection occurs, especially in patients with multiple tumors (Kato *et al.*, 1999; Guo *et al.*, 2008). Our patient had a solitary nodule, which did not recur 2 years post surgery upon follow up. The younger age of our patient and the location of the tumor, both being rare presentations make it an interesting case.

In conclusion, although very rare, pulmonary hamartomas can occur in young age with the central location. Clinically, the presentation is usually non-specific and the diagnosis is mostly incidental while investigating other conditions. Surgical resection is the mainstay of treatment for this condition. There is a need to increase awareness among clinicians on hamartomas to improve early treatment for this rare disease in the adolencents.

Conflict of Interests

The authors declare that they have no conflict of interests.

Consent

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Written informed consent was obtained from the patient for publication of this paper and accompanying images.

Authors' Contribution

BM, JRM SBK: managed the patient, BM, HR, JRM, JD and SBK collected the clinical information and wrote the manuscript. All authors critically revised the manuscript and approved the final draft

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