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A rare case of pulmonary hyalinizing granuloma with calcification in a 5 year old boy

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ABSTRACT

Pulmonary hyalinizing granuloma (PHG) is a rare benign pulmonary nodular lesion of unknown etiology. We present a case of a 5-year-old boy who was found to have a chest mass while being evaluated for abdominal pain. He underwent a CXR and CT scan that showed popcorn calcifications in the right posterior mediastinum and within the hilum of right lung. These lesions were suspicious for benign calcified lymph nodes and follow-up chest CT after 3.5 months showed no interval changes in the calcified mediastinal masses. Extensive testing ruled out infectious diseases and malignancies. Given the unknown etiology of the lesions, he underwent VATS biopsy that demonstrated a nodular lesion characterized by a peripheral rim of fibrous tissue and central zone of necrosis and calcification, findings consistent with hyalinizing granuloma. PHG is extremely rare in pediatric age group. Although diagnosis of this condition is made by radiological and histopathological findings, it is important to rule out other causes of chest masses. Most of the patients usually have good prognosis with this rare disorder.

Pulmonary Hyalinizing Granuloma (PHG) is a rare, benign pulmonary nodular lesion of unknown etiology and is characterized pathologically by whorled deposits of lamellar collagen. First described by Engleman et al. [1] in 1977, there have been many descriptions and case reports of this disease; however, most of the case reports have been in adults with secondary associations with TB, fungal infections or carcinomas. The youngest case was an adolescent child [2]. We present the youngest child to date of an incidentally discovered case of PHG.

1. Case report

A 5-year-old Caucasian male with past history of ADHD, anxiety and history of constipation presented to urgent care for abdominal pain. He underwent abdominal radiography that showed a normal bowel pattern, but was incidentally noted to have calcified nodules in the right lung.

The child had no history of fevers, night sweats or weight loss. His mother noted that he had generalized fatigue over the previous month, but otherwise was in his usual state of health. Approximately a year before presentation, he exhibited a month-long URI-type illness predominated by cough. His history was also significant for being exposed to an abandoned chicken coop but he did not have any direct contact with farm animals or birds. His travel had been limited to southeast United States. On physical exam, he was a healthy child without any respiratory findings.

A repeat chest radiograph confirmed the previous findings of a right parasternal calcified lesion measuring 1.7 × 1.6 cm. He underwent chest CT scan without contrast which showed popcorn calcification in the right hilum and azygoesophageal groove, suspicious for benign calcified lymph nodes. Given the benign appearance of lesion, the child was observed and a follow up CT scan was performed at 3 months. The chest CT showed no change in either the mediastinal or hilar lymph nodes, but additional lesions were identified within the left lower lobe and a new lesion was noted within the right middle lobe (Figs. 1–3).

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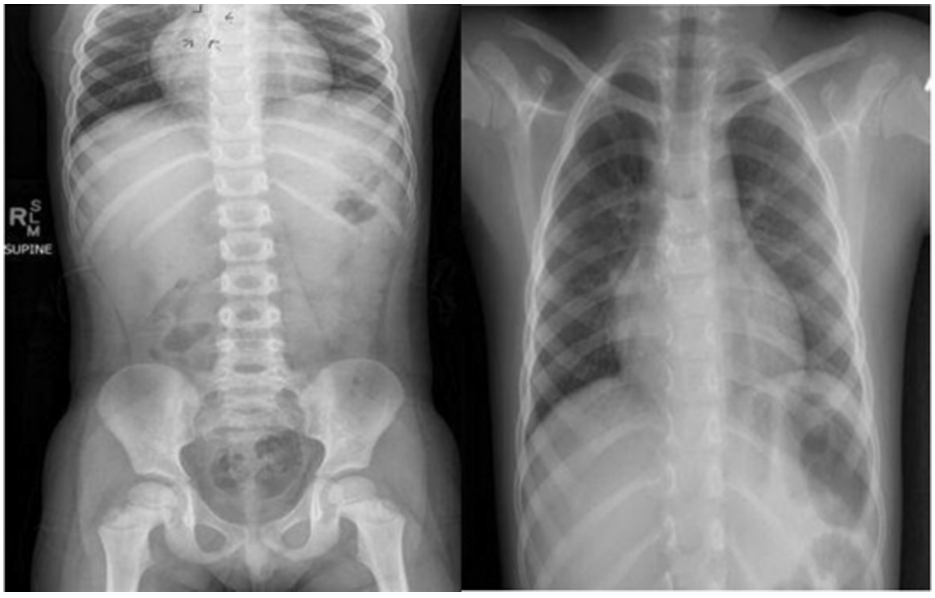

Fig. 1. Incidental finding of pulmonary calcification.

His workup ruled out histoplasmosis, sarcoidosis, hamartomas, and malignancy. Urine histoplasma antigen and serum histoplasma antibodies were negative. Serum ACE level was normal. Other routine labs, including complete blood count, electrolytes, and coagulation were normal. H. Capsulatum antibody immunodiffusion was negative.

In order to diagnose the lesion, a VATS biopsy was performed. The mass was noted to be in the retropleural space at approximately the level of the fourth intercostal space. Approximately 75% of the mass was removed. The gross findings showed a $2.0 \times 1.2 \times 1.0$ cm tan portion of soft tissue with the cut surface revealing a 1.5 cm cavity containing an abundant amount of

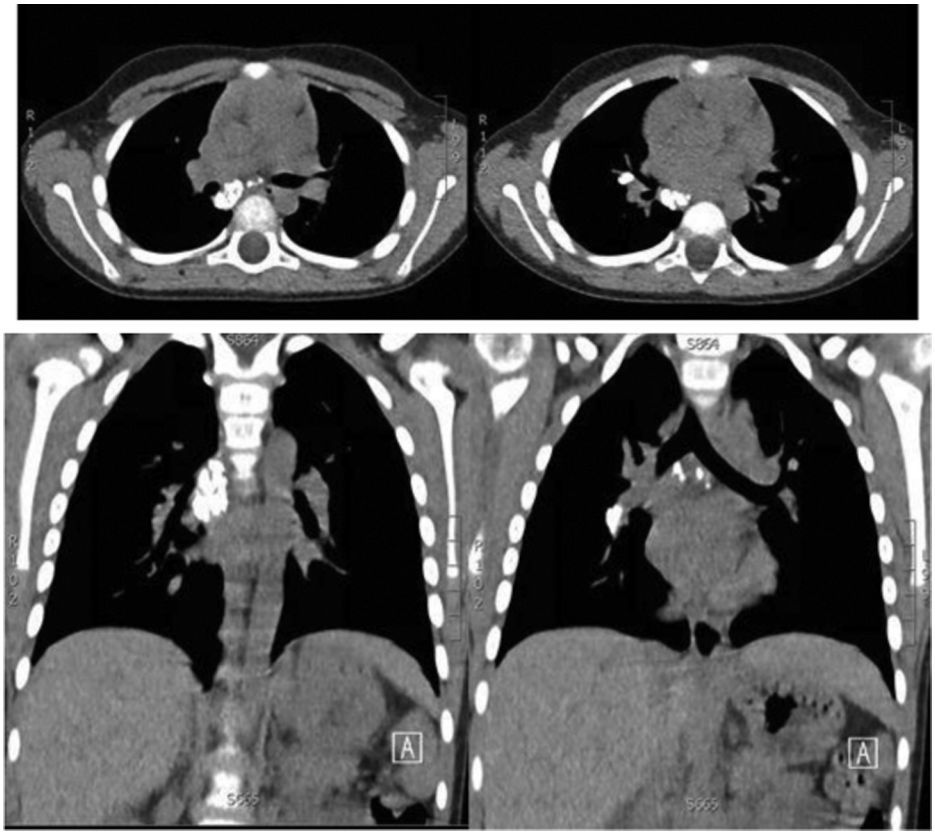

Fig. 2. CT scan findings of the perihilar mass showing calcification.

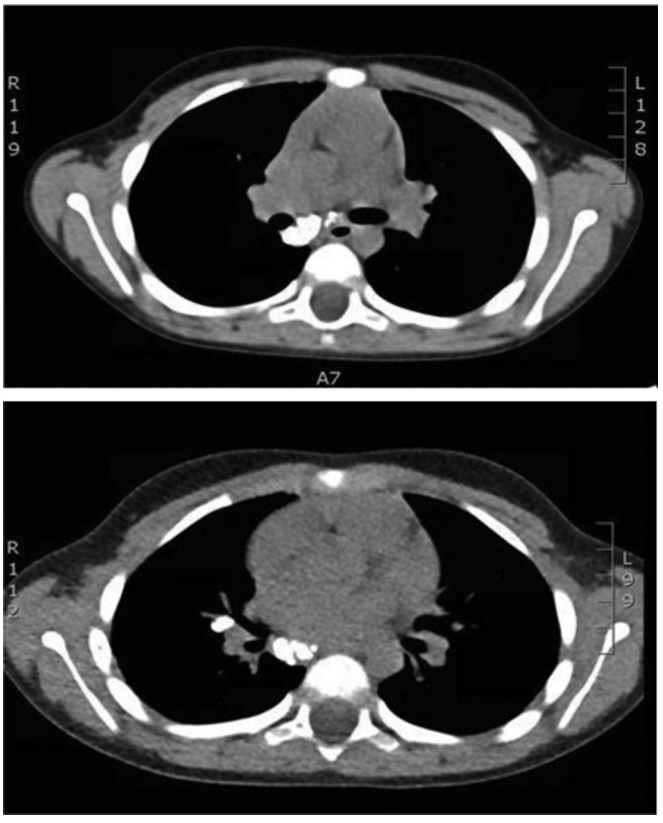

Fig. 3. CT scan follow-up findings after 3.5 months.

tan-white material. Microscopic examination showed nodular lesion characterized by a peripheral rim of mature fibrous tissue and a central zone of necrosis and calcification (Fig. 4). No acid fast bacilli were demonstrated with Kinyoun stain. No fungus identified with Grocott-Methenamine silver stain. Gram stain and cultures were negative for aerobic and anaerobic organisms. Subsequent extensive rheumatology and allergy evaluations were negative.

2. Discussion

Pulmonary hyalinizing granuloma (PHG) was first described by Engleman et al. in 1977 and over the last three and a half decades, there have been many publications describing various associations and clinical presentations. Most commonly, PHGs occurs in middle-aged adults with mean age of 45 years, without gender predominance [1,3,4]. To date, the youngest patient was a 15-year-old male who had associated retroperitoneal fibrosis [2].

The etiology of PHG appears to have an association with infectious or autoimmune pathology. In the initial report by Engleman et al. [1], many patients had prior history of TB, or were complicated by Sclerosing mediastinitis, retroperitoneal fibrosis or amyloidosis [5]. Half of the reported patients with PHG have an exposure to Histoplasma or mycobacterium [6]. Similarly, up to 60% of patients with PHG have evidence of a specific underlying autoimmune disorder [7]. Several other case reports demonstrate associations of PHG with immunologic processes such as elevated ANA, Rheumatoid factor, circulating immune complexes, lymphomas, Castleman's disease, Reidel's thyroiditis, Aspergillosis, and multiple sclerosis [3,5]. Additionally, inflammatory myofibroblastic tumor (IMT) has been associated with PHG. PHG and IMT occur in different age groups. PHG occur most commonly in middle aged adults whereas IMT occurs predominantly in younger age group. IMT is the most common endobronchial mesenchymal tumor in children and most common primary lung tumor in pediatric age group [5].

Various clinical presentations have been described in adult literature. Young et al. described the first pediatric case report of PHG associated with retroperitoneal fibrosis [2]. In general, the clinical presentation of PHG can vary. In 25% of patients, the disease is asymptomatic and the nodules are incidentally noted (Pinckard) [2]. Symptoms such as chest pain, dyspnea or hemoptysis in association with multiple bilateral parenchymal nodules are also described [2].

Diagnosis is made by radiological and histopathological findings. Chest radiography and CT usually identifies the lesion but are non-specific in their presentation. Patients with PHG typically reveal nodules that range in size from 0.2 cm to 15 cm with mean size of 2 cm [2]. The nodules have variably circumscribed borders and sometimes have an ill-defined cotton-ball-like appearance [6]. Most of the lesions present as multiple nodules [4]. Presentation as a solitary nodule is usually rare [5,8]. The lesion can be situated in the lung parenchyma or subpleura [4,9]. Usually they grow slowly or do not grow at all, and spontaneous regression has been described [4,10].

Definitive diagnosis of PHG can only be achieved through tissue biopsy [3,5,11]. The lesions are described as well-circumscribed nodules with the center of nodule being occupied by lamellae of hyalinized collagen. In PHG, the deposition pattern of lamellar collagen is diagnostic and is characterized by ropy whorled collagen bundles separated by clear spaces [3].

Engleman described the prognosis of the condition as very good or status quo, with nodules growing very slowly or even with

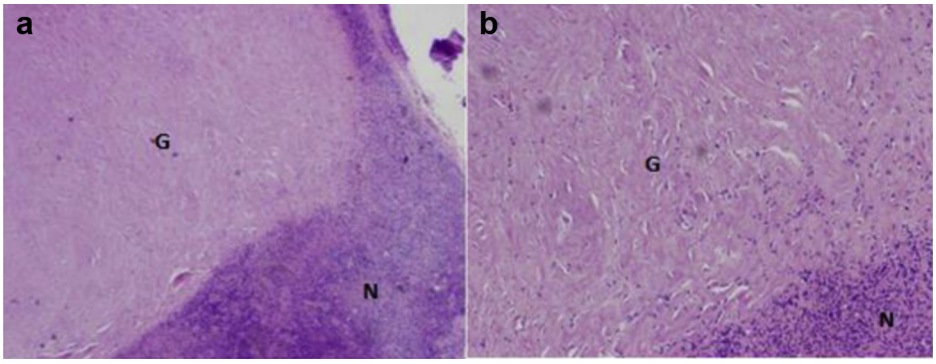

Fig. 4. Histological findings of the tissue sample (a) 16× and (b) 40× view of the mass showing hyalanized granuloma (pink, G) within the lymph node (blue, N). Noted are areas with peripheral fibrous tissue and central zone of necrosis and calcification.

spontaneous regression possibly noted [1,12,13]. Up to 30% of patients with PHG have progressive disease with enlarging nodules and increased dyspnea [7,14]. Schlosnagle et al. reported that nodules did not change after treating with prednisolone [3,15].

Extrapulmonary involvement is seen in approximately 20% of reported cases and when the nodules are located near the hilum or mediastinum, fibrosing mediastintitis may be prone to develop [4,7,9]. This is the most common complication that is developed in patients with PHG.

3. Conclusion

We present the youngest case to date of pulmonary hyalinizing granuloma. PHG is a rare fibrosclerotic inflammatory disease with extremely rare presentation in the pediatric age group. Patients who present with pulmonary nodules should be thoroughly evaluated for other inflammatory disorders, tumors, infectious diseases. These children will require tissue biopsy and close observation. Most of the patients have good prognosis. Histopathological evaluation combined with radiological tests guide in surgical evaluation and management of such patients.

Conflict of interest

Authors have no relevant conflicts of interest to disclose.

References

[1] Engleman P, Liebow AA, Gmelich J, Friedman PJ. Pulmonary hyalinizing granuloma. Am Rev Respir Dis 1977 Jun;115(6):997–1008.
[2] Young AS, Binkovitz LA, Adler BH, Nicol KK, Rennebohm RM. Pulmonary hyalinizing granuloma and retroperitoneal fibrosis in an adolescent. Pediatr Radiol 2007 Jan;37(1):91–5.
[3] Anazawa Y, Nagai H, Motomiya M, Isawa T, Saito Y, Takahashi T, et al. A case of pulmonary hyalinizing granuloma. Tohoku J Exp Med 1992 May;167(1):39–45.
[4] Na KJ, Song SY, Kim JH, Kim YC. Subpleural pulmonary hyalinizing granuloma presenting as a solitary pulmonary nodule. J Thorac Oncol 2007 Aug;2(8):777–9.
[5] Xu ZL, Bethune D, Manos D, Foyle A, Henteleff H, Johnston M, et al. Pulmonary hyalinizing granuloma. Beijing Da Xue Xue Bao 2009 Aug 18;41(4):463–8.
[6] Patel Y, Ishikawa S, MacDonnell KF. Pulmonary hyalinizing granuloma presenting as multiple cavitary calcified nodules. Chest 1991 Dec;100(6):1720–1.
[7] Yousem SA, Hochholzer L. Pulmonary hyalinizing granuloma. Am J Clin Pathol 1987 Jan;87(1):1–6.
[8] Eschelman DJ, Blickman JG, Lazar HL, O'Keane JC, Schechter M. Pulmonary hyalinizing granuloma: a rare cause of a solitary pulmonary nodule. J Thorac Imaging 1991 Apr;6(2):54–6.
[9] Esme H, Ermis SS, Fidan F, Unlu M, Dilek FH. A case of pulmonary hyalinizing granuloma associated with posterior uveitis. Tohoku J Exp Med 2004 Sep;204(1):93–7.
[10] Popat S, Nicholson AG, Fisher C, Harmer C, Moskovic E, Murday VA, et al. Pulmonary masses presenting 11 years after abdominal surgery. Respiration 2004 May-Jun;71(3):295–7.
[11] Guccion JG, Rohatgi PK, Saini N. Pulmonary hyalinizing granuloma electron microscopic and immunologic studies. Chest 1984 Apr;85(4):571–3.
[12] Gans SJ, van der Elst AM, Straks W. Pulmonary hyalinizing granuloma. Eur Respir J 1988 Apr;1(4):389–91.
[13] Ren Y, Raitz EN, Lee KR, Pingleton SK, Tawfik O. Pulmonary small lymphocytic lymphoma (mucosa-associated lymphoid tissue type) associated with pulmonary hyalinizing granuloma. Chest 2001 Sep;120(3):1027–30.
[14] Ramirez J, Mehta JB, Taylor RA, Byrd Jr RP, Roy TM. Symptomatic pulmonary hyalinizing granuloma. South Med J 1998 Sep;91(9):867–9.
[15] Schlosnagle DC, Check IJ, Sewell CW, Plummer A, York RM, Hunter RL. Immunologic abnormalities in two patients with pulmonary hyalinizing granuloma. Am J Clin Pathol 1982 Aug;78(2):231–5.