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**Case Report**

# A child presenting with bullous emphysema

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## ABSTRACT

**Background.** Placental transmogrification of the lung (PTL) is a clinical spectrum varying from asymptomatic to severe pulmonary impairment; such as recurrent pneumothorax, bronchopneumonia, respiratory distress syndrome and chronic obstructive airway disease. PTL usually presents as a bullous lesion, and rarely can appear in nodule or cyst formation on chest imaging. PTL with giant bullous emphysema has a male preference, is more commonly unilateral and mostly affects one lobe, but can rarely involve more than one lobe.

**Case.** Here we report a 13-year-old boy presenting with bullous emphysema and coexisting with a borderline testicular tumor. He had no complaints of cough, sputum, or shortness of breath. He had a past medical history of pneumonia five years ago. In order to elucidate the underlying lung pathology, a wedge lung biopsy was performed and the patient was diagnosed with PTL. Scrotum ultrasonography was performed because of hydrocele in both testes, and bilateral epididymal cysts with papillary solid projections were reported. Pathological examination of the epididymal tumor revealed a "Mullerian type borderline epithelial neoplasm" which is an analogue of the ovarian serous borderline tumor.

**Conclusions.** In conclusion, we reported the youngest PTL case in the literature, a rare disease with unknown pathophysiology, presenting as bullous emphysema and coincidental Mullerian type borderline epithelial neoplasm. It is important to diagnose placental transmogrification of the lung in a child with bullous emphysema because compared to other cystic lung diseases it is a benign disease and if no additional malignity exists, lobectomy or pneumonectomy is the cure for the disease.

**Key words:** placental transmogrification, child, bullous emphysema, Mullerian type borderline epithelial neoplasm, hydrocele.

Placental transmogrification of the lung (PTL) which was named after the resemblance to immature placental villous structures was first described by McChesney in 1979.[1] Placental transmogrification can be asymptomatic, however severe pulmonary impairment; such as recurrent pneumothorax, bronchopneumonia, respiratory distress syndrome and chronic obstructive airway disease can be seen. Radiological differential diagnosis of PTL includes pneumonia, bronchogenic cyst, alveolar adenoma, sclerosing hemangioma, congenital pulmonary airway malformation, hamartomas.[2] PTL frequently appears as a bullous lesion. However, it can rarely appear in nodule or cyst formation on chest imaging.[3] PTL with giant bullous emphysema has a male preference, is more commonly unilateral and mostly affects one lobe, but can rarely involve more than one lobe.[2]

Here we report a 13-year-old boy presenting with giant bullous emphysema and coexisting with a borderline testicular tumor with review of the literature. The young age of our patient and the rarity of this pathological diagnosis and the coexisting testicular tumor makes this case unique.

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## Case Report

A 13-year-old boy was admitted to our pediatric pulmonology outpatient clinic due to a chest deformity recognized by his parents. He had no complaints of cough, sputum, or shortness of breath. He had a past medical history of pneumonia five years ago, in which he was hospitalized in a different facility and treated for, and discharged without any morbidities. During this hospitalization a chest computed tomography (CT) was performed indicating ground glass opacities and interlobular septal thickening, no further work up was performed. In the same year, the patient had an endovenous laser ablation procedure due to Vena Saphena Magna ectatic venous formation. For the following five years he had no complaints, and was lost to follow up. On admission to our hospital in 2019, the physical examination revealed that his weight and height were within the normal percentile, transcutaneous oxygen saturation was 96% at room air, he had an asymmetrical chest wall with protrusion of the left chest wall, decreased lung sounds on the right lower lobe, levoscoliosis at the level of thoracic vertebra, and bilateral hydrocele in testis. He had no signs of hemihypertrophy. Spirometry revealed a restrictive pattern (Vital capacity (VC): 53% forced VC (FVC): 50%, forced expiratory volume in 1 Second (FEV1): 50% FEV1/FVC: 83 forced expiratory flow at 25-75% of the pulmonary volume (FEF25/75): 45%). Chest CT was performed displaying fibrotic bands, septal thickening, emphysematous regions, honeycombing and air trapping mainly localized to the right lower and middle lobe and a leftward mediastinal shift (Fig. 1). A progression was observed at the right lung parenchyma within five years. Because of the localized nature of the pulmonary disease and a history of venous pathology at the right leg, we wanted to exclude venous lymphatic drainage diseases, so we performed a lymphoscintigraphy in which no lymphatic pathology was found. Scrotum ultrasonography was performed because of hydrocele in both testes, and bilateral epididymal cysts with papillary solid projections were reported (Fig. 2). Bilateral tumor was successfully resected by pediatric surgeons.

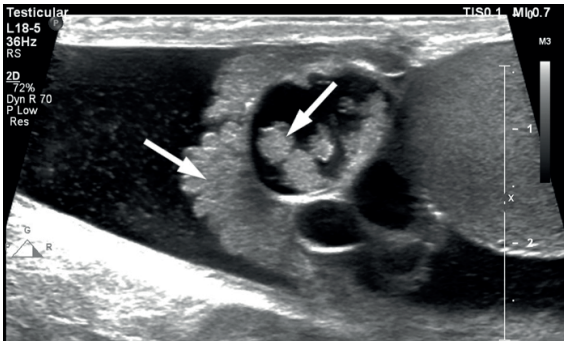


**Fig. 2.** Ultrasonography image demonstrates a cystic neoplasm with papillary projections of the epididymis (arrows). Note the normal testis parenchyma.

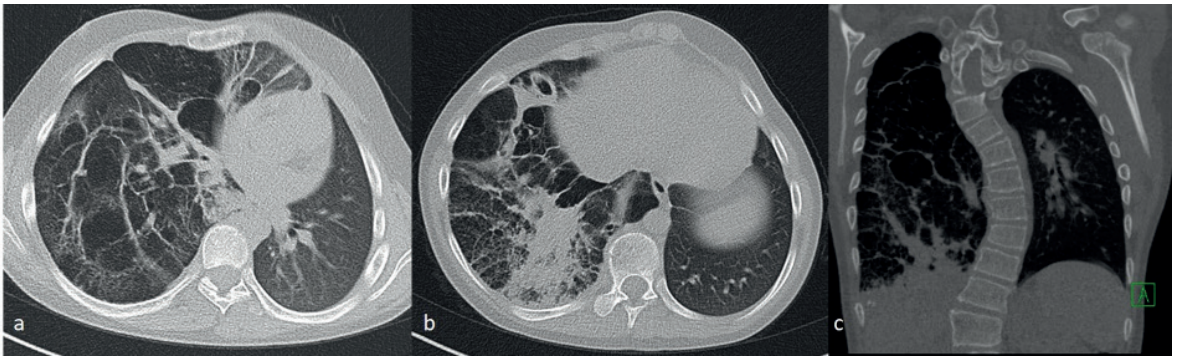


**Fig. 1. A, B)** Axial chest CT images show, fibrotic bands, septal thickening, bullous/emphysematous changes, air trapping and leftward mediastinal shift, honeycombing mainly localized to right lower lobe. **C)** Coronal reformatted image demonstrates levoscoliosis of the thoracic spine.

Pathological examination of the epididymal tumor revealed a "Mullerian type borderline epithelial neoplasm" which is an analogue of the ovarian serous borderline tumor (Fig. 3A). The tumor was located in the right epididymis, causing a cystic dilatation with numerous intra-cystic blunt papillae lined by ciliated stratified columnar cells with mild cytological atypia. The cyst wall had a variable amount of fibrous tissue without any invasion or necrosis detected. Immunohistochemical studies performed for ER, PR and CK7 revealed positivity in neoplastic cells, while very focal CD10 expression was detected only in the apical part of the cells. D2-40 and calretinin were negative, excluding a tumor with mesothelial origin. Ki-67 proliferation index varied from 2-3% to 10% in the lowest and highest areas (mean 5%), respectively. Considering both morphological and immunohistochemically findings, the biopsy confirmed a diagnosis of borderline serous tumor with a Mullerian origin. Because of the intact surgical margin, and negative tumor markers (Ca-125) total resection of the tumor was thought to be curative and clinical follow up was planned by pediatric oncology.

In order to elucidate the underlying lung pathology, a wedge lung biopsy was performed. The histopathologic examination showed numerous irregular cystic/emphysematous parenchymal areas accompanied by papillary structures surrounded by flattened alveolar pneumocytes, morphologically resembling placental chorion villi (Fig. 3B), especially at low power view magnifications. The patient was diagnosed with PTL. Because of the bullous/

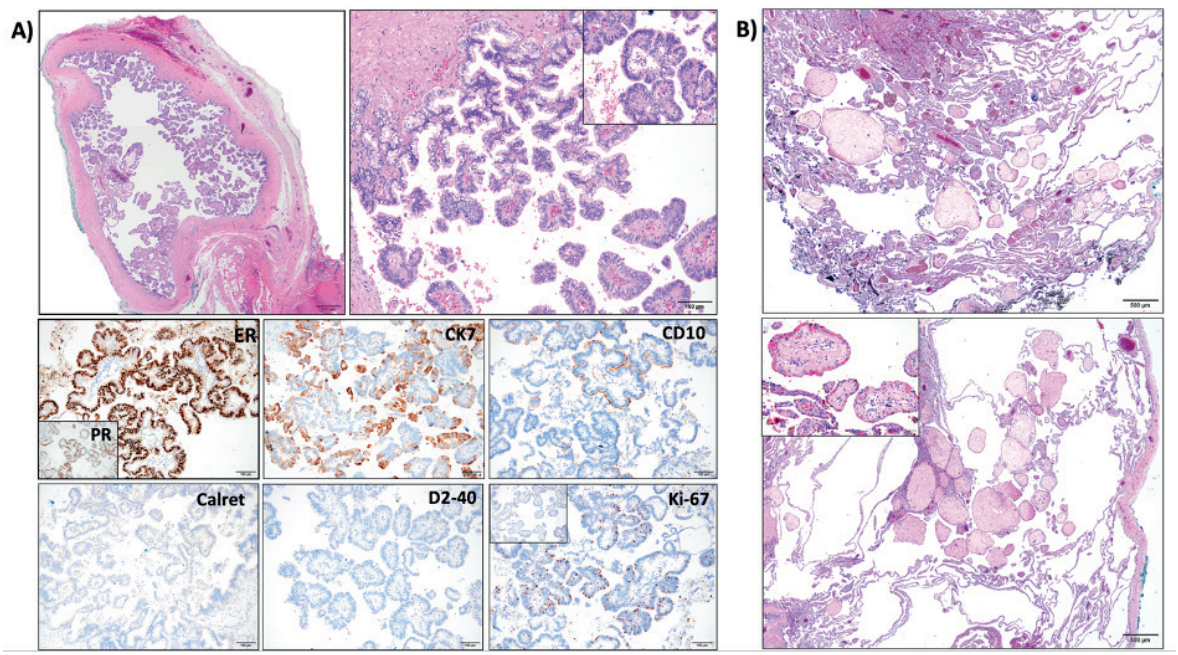


**Fig. 3. A)** Low power view of the epididymal cystic neoplasm having papillary projections towards the lumen (upper left: HE, x20); papillary projections showing «medusa head» micropapillary pattern (upper right: HE, x200); intra-cystic blunt papillae lined by stratified epithelial cells with mild cytologic atypia (inlet: HE, x200 ); summary of the immunohistochemical findings suggesting a Mullerian origin rather than a mesothelial one; ki-67 with high and low areas (lower pictures: IHC, x100 for each, respectively). **B)** Low power view of the pulmonary placental transmogrification within large cystic emphysematous areas (upper: HE, x20); pulmonary parenchyma includes cystic lesions with variable amounts of intra-cystic papillary proliferations (lower: HE, x20); papillary structures, which morphologically resemble mature chorionic villi, contain congested capillaries and are surrounded by alveolar pneumocytes (inlet: HE, x200).

emphysematous changes which severely compressed the normal lung tissue and leftward mediastinal shift, right middle and right lower lobectomy were planned. During the procedure the right upper lobe was not inflated after resection of the right middle and lower lobe, so right pneumonectomy was performed. The pathological result was compatible with placental transmogrification.

During the post-operative period, the patient had no complaints except for exertion dyspnea during heavy physical exercise. Spirometry revealed a restrictive pattern (VC: 68% FVC: 67%, FEV1: 72% FEV1/FVC: 88 FEF25/75: 74%). The patient was referred to orthopedic surgeons and no surgical intervention was planned for scoliosis.

The patient and the caregiver gave written consent for the publication of this case.

## Discussion

PTL is a rare disease. As of 2019, less than 40 adult patients with PTL were defined in the literature. Until 2017, there were no children diagnosed with PTL in the literature. This is the third case of a child in the literature that has been reported so far. The young age of our patient and the rarity of this pathological diagnosis and coexisting testicular tumor makes this case unique. In this case report, we tried to emphasize the importance of differential diagnosis in a patient presenting with giant bullous emphysema who was nearly asymptomatic when he was first admitted to our unit. It is important to diagnose PTL in a child with giant bullous emphysema because compared to other cystic lung diseases it is a benign disease and if no additional malignity exists lobectomy or pneumonectomy is the cure for the disease.

Knowledge concerning PTL in children is limited. To date, there are only two case studies reporting PTL in children, and in both of the cases giant bullous emphysema cases were restricted to one side of the lung and more than one lobe was affected.[4,5] In both of the cases the patients were 14-year-old males, presenting with back pain, and no other significant physical findings. One child had chest CT findings in past years where pneumonia or bleb formation could not be differentiated, both patients were cured by lobectomy of the right middle lobe and right lobe, intervention. There was no comorbidity in both patients. In contrast, our case was diagnosed with testicular Mullerian type borderline epithelial neoplasm - the analogue of ovarian serous borderline epithelial tumor-, at the same time interval. The borderline serous tumor one of the epithelial paratesticular tumors with Müllerian characteristics was first reported by Young and Scully in 1986.[6] To date only four children with testicular ovarian epithelial tumor, serous borderline have been reported. A painless mass with hydrocele was detected in all of these patients. In our case, Ki67 proliferation index was 5% which is consistent with the literature where Ki67 proliferation index ranges between 1.3-10%.[7,8] However, none of the cases with testicular Mullerian type serous borderline epithelial tumor, had a coexisting lung pathology. Because of the papillary structures, it can be tricky to differentiate a metastatic papillary neoplasm from PTL's papillary figures in a low power view. On the other hand, cytologic details of the epithelial component of the serous tumor can be easily distinguished from single layered bland pneumocytes of PTL. Besides, another major histopathological difference between these two lesions - architectural simplicity of PTL and "medusa head" pattern seen in serous tumor- was proof that they are not linked with each other. Therefore, we considered these two diseases co-existing in our case as two different clinical entities.

PTL usually presents as a bullous lesion, and rarely can appear in nodule or cyst formation on chest imaging.[2] In our case, in addition to bullous/emphysematous changes, other radiological findings such as: air trapping, fibrotic bands, septal thickening, mediastinal shift and honeycombing localized to the

unilateral lung were also present. We believe these additional radiological findings can be explained by the delayed diagnosis because the patient was asymptomatic and did not apply to a hospital for almost five years.

Because of the extreme rarity of PTL the pathogenesis remains unknown. However, Narula et al.[9] suggested that the mechanism of disease can range from lymph vascular proliferation in the setting of an emphysematous lung to congenital malformation, which can both be the case in our patient as he had abnormal lymphovascular proliferation history of the right leg and existing lung lesion on chest CT when he was 8 years old. One of the other possible mechanisms for PTL development is increased fat tissue expression inside the villi arising from lipomatosis.[3] In one case series, it was stated that emphysematous changes can be caused by the primary inciting event: interstitial clear cell proliferation.[2]

In our case, the patient had bullous/emphysematous changes which severely compressed the normal lung tissue and leftward mediastinal shift. Multiple (right middle and lower lobe) lobectomy was planned however after the multiple lobectomies, the right upper lobe was not inflated, so right pneumonectomy was performed. Ma et al.[10] stated in their study that among 33 adult patients the most common choice for operation was lobectomy, but pneumonectomy was performed in 7 patients. The authors concluded that the size and extension of the lesions are associated with the type of surgery. They highlighted that when giant bullae were present which severely compressed the normal lung tissue multiple lobectomy or pneumonectomy had to be performed.[10]

In conclusion, we report the youngest PTL case in the literature, a rare disease with unknown pathophysiology, presenting as giant bullous emphysema and coincidental Mullerian type borderline epithelial neoplasm. PTL should be kept in mind in a child presenting with unilateral emphysema because it is a curable disease, unlike most cystic lung diseases of childhood.

## Ethical approval

The patient and the caregiver gave written consent for the publication of this case.

## Author contribution

The authors confirm contribution to the paper as follows: study conception and design: BO DAT, MÜ, HNÖ, EN, NE, TS; draft manuscript preparation: BO, DO, EY, DD, UO, NK. All authors approved the final version of the manuscript.

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## Conflict of interest

The authors declare that there is no conflict of interest.

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