

## PLEUROPULMONARY BLASTOMA PRESENTING AS A COMPLICATED PLEURAL EFFUSION

J. O'Brien, D. Rea, R. Hayes<sup>1</sup>

**Pleuropulmonary blastoma (PPB) is a rare tumour of mesenchymal cells. We present a case of PPB in a child, which presented to the emergency department with an extensive pleural effusion. We discuss the radiological features, pathology, classification and treatment of this condition. This case reiterates the importance of considering this diagnosis prior to surgical intervention, to improve the long term prognosis of this aggressive disease.**

**Key-word:** Blastoma.

### Case report

A 3-year-old previously healthy girl was referred to the emergency department with a 2-day history of increasing dyspnoea and drowsiness. Laboratory investigations revealed an elevated white cell count of  $20 \text{ cells/mm}^3$ , with normal haemoglobin and platelet levels. ESR was elevated at 110 mm/hour. A chest x-ray revealed extensive opacification of the right hemithorax with minimal aerated lung in the right apex. There was marked mediastinal shift to the left and patchy left lower lobe consolidation. There was also a small air pocket projected to the right of the midline which was suggested to be a pneumatocele (Fig. 1). An ultrasound of her chest revealed that the right lung was abnormally echogenic with multiple fluid-filled cystic spaces and was surrounded by a large collection of septated fluid which suggested empyema (Fig. 2). The diagnosis of underlying lung abscess was proposed. A subsequent Computed Tomography (CT) thorax was performed (Fig. 3) which confirmed mediastinal shift to the left, with a thick rind of peripulmonary fluid. Furthermore, the right lower lobe parenchyma appeared abnormal with a mixed solid and cystic appearance. The patient was referred for thoracotomy, with a histopathological diagnosis of pleuropulmonary blastoma.

### Discussion

Pulmonary blastoma is a rare aggressive neoplasm which usually presents as a well-defined lung lesion or pleural effusion and

accounts for 0.25-0.5% of lung malignancies. These tumours are composed of malignant immature epithelial or mesenchymal cells which may bear a resemblance to early embryological lung tissue. They can be classified into three subgroups: Classic pulmonary blastoma (PB), well-differentiated fetal adenocarcinoma (W DFA) and pleuropulmonary blastoma (PPB) (1).

Classic pulmonary blastoma is the most common subtype and often presents with non-specific respiratory symptoms with two peak age incidences in the first and fourth decades of life. Chest x-rays reveal a solitary lung mass or nodule, however 40% may be diagnosed coincidentally.

W DFA usually affects adults with a history of smoking and may be mistaken for bronchogenic carcinoma. Pleuropulmonary blastoma is a distinct tumour of childhood and can be classified into three subtypes depending on the histological appearance. Type I is a purely cystic tumour, type II has mixed solid and cystic components and type III is a solid tumour (2). There have been reports of a better prognosis associated with type I disease in comparison with Types II and III, however this has not proven to be statistically significant and may be due to the lack of data and the rarity of this tumour.

On CT, PPB appears as a mixed solid and cystic lesion with a necrotic centre and variable contrast enhancement, or a persistent pleural effusion and can cause complete opacification of a hemithorax, with co-existing mediastinal shift. Differential diagnosis depends on the presentation, however, conditions such as congenital cystic adenomatoid malformation (CCAM)



*Fig. 1.* — Chest X-ray demonstrating extensive opacification of the right hemithorax with marked mediastinal shift to the left and patchy left lower lobe consolidation. There was an area of aerated lung at the right apex and also a small air pocket projected to the right of the midline.

*From:* 1. Dept. of Radiology, Our Lady's Hospital for Sick Children, Crumlin, Dublin, Ireland.

*Address for correspondence:* Dr J. O'Brien, M.D., Dept. of Radiology, Our Lady's Hospital for Sick Children, Dublin, Ireland. E-mail: juliemobrien@gmail.com



Fig. 2. — Longitudinal ultrasound scan over right lung demonstrating echogenic parenchyma with septated fluid.



Fig. 3. — Axial post-contrast CT scan of the thorax confirms a mixed solid/cystic mass in the right hemithorax with surrounding peripulmonary fluid. There is also significant shift of the mediastinum to the left.

should also be considered and it can be difficult to distinguish this from PPB type I (3). It is also important to note that, these lesions can often arise in association with congenital lung lesions such as the previously mentioned CCAM (4). In the case of an opaque hemi-thorax with an associated rib lesion, consideration should also be given to lymphoma or Ewing's sarcoma. There are no specific genetic markers yet identified despite extensive investigation (5), however 25% cases are associated with familial malignancies including sarcomas, medulloblastomas, lymphoma, leukaemia and PPB.

Patients are often commenced on treatment for an empyema with a poor response.

Surgery is the mainstay of treatment, with a combination of

chemotherapy and radiotherapy, however it is an aggressive tumour with a relatively poor prognosis, which increases with tumour masses larger than 5 cm at diagnosis.

Unfortunately, a pre-operative diagnosis is rarely made, and the diagnosis is usually seen retrospectively on histopathological analysis of the surgical specimen which has often not achieved definitive surgical margins. Since an adequate resection is required to prevent tumour seeding, a pre-operative consideration should be given to this diagnosis the presence of these features to ensure a better prognosis.

#### References

1. Walker R., Suvarna K., Matthews S.: Pulmonary blastoma: presentation of
2. Priest J.R., McDermott M.B., Bhatia S., Watterson J., Mannivel J.C., Dehner L.P.: Pleuropulmonary blastoma: a clinicopathologic study of 50 cases. *Cancer*, 1997, 80 (1): 147-161.
3. Orazi C., Inserra A., Schingo PM., De Sio L., Cutrera R., Boldrini R., Malena S.: Pleuropulmonary blastoma, a distinctive neoplasm of childhood: report of three cases. *Pediatr Radiol*, 2007, 37: 337-344.
4. Naffaa L., Donnelly L.: Imaging findings in pleuropulmonary blastoma. *Pediatr Radiol*, 2005, 35: 387-391.
5. Taube J.M., Griffin C.A., Yonescu R., Morsberger L., Argani P., Askin F.B., Batista D.A.: Pleuropulmonary blastoma: cytogenetic and spectral karyotype analysis. *Pediatr Dev Pathol*, 2006, 9: 453-461.