Prenatal detected retroperitoneal pulmonary sequestration with elevated serum levels of CA 19-9 – Case report and review of the literature

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A B S T R A C T
We present a case of prenatal detected retroperitoneal pulmonary sequestration (RPS) with an interesting finding. Immunohistochemical staining of the specimen indicated that the elevated carbohydrate antigen (CA) 19-9 serum level was very likely caused by the sequestration. A review of the literature was made. RPS is an infrequent congenital malformation, which can be detected by routine obstetric sono- graphic screening during prenatal life. It can be misdiagnosed as neuroblastoma. The differential diagnosis can be tricky, which we hope our finding can help with. The management remains controversial.

1. Case report

A left suprarenal fetal mass was discovered at 5 months’ gestation on routine prenatal ultrasound scan of a 31-year-old gravida 1, para 0 woman. The mass was hyperechoic, measured 2.3 \times 1.8 \text{cm} at 28 weeks’ gestation (Fig. 1A). An magnetic resonance imaging (MRI) scan at the same time showed the lesion was hyperintense on T2-weighted (Fig. 1B). To our knowledge, the most possible diagnosis was either neuroblastoma or adrenal hematoma. We applied the strategy of “wait and see.” Three repeat ultrasound scans at 37 weeks’ gestation, 1 week after born (full-term normal delivery at 38 week’s gestation, mother and child didn’t show any abnormal symptoms and physical signs), 4 weeks showed the persistence of the lesion. Further evaluation was performed because of the suspicion of malignancy at 6 weeks after birth. The urinary catecholamines assay, AFP, \(\beta\)-HCG were all normal. The carbohydrate antigen (CA) 19-9 was 53 \text{ng/ml}, when the normal upper limit was 38 \text{ng/ml}. The detection for neuroblastoma minimal residual antigen (CA) 19-9 was 53 \text{ng/ml}, when the normal upper limit was 38 \text{ng/ml}. The detection for neuroblastoma minimal residual antigen (CA) 19-9 serum level was very likely caused by the sequestration. A review of the literature was made. RPS is an infrequent congenital malformation, which can be detected by routine obstetric sono- graphic screening during prenatal life. It can be misdiagnosed as neuroblastoma. The differential diagnosis can be tricky, which we hope our finding can help with. The management remains controversial.

2. Discussion

Pulmonary sequestration (PS) is a segment of lung that has no communication with the normal bronchial tree and receives blood supply from systemic arteries. The incidence is about 0.15—1.8% [1]. Extralobar pulmonary sequestrations (ELS), which account for 25% of prenatal detected lung lesions [1], are masses of lung parenchyma that have a distinct pleura. About 10—15% of ELS will occur in retroperitoneal sites [1]. RPS is believed to result from the formation of an accessory lung bud on special level of the foregut [1]. In countries where routine prenatal ultrasound has been carried out for a period of time, RPS seems the third common suprarenal localized masses diagnosed during the perinatal period [2].

In our case, the diagnosis of RPS did not occur to us until the pathology result due to lacking of awareness. After reviewing the literature, we found that our case shared most characteristics of prenatal detected RPS: gestational age at detection could be as early as third month; It was typically located in the left suprarenal area; ultrasound study usually showed hyperechogenic lesion, with or
without cystic components; It was usually a hyperintense lesion on T2-weighted MRI images, and was a heterogeneous, hypodense mass that increase density with intravenous contrast on CT scan [3]; Prenatal and postnatal natural history of RPS is usually uneventful [4]. It was a pity that the information about the feeding systemic arterial vessel(s), the most distinctive characteristic of RPS, remained unclear in our case. Without awareness, neither did we take measures like high resolution CT or angio-MRI to demonstrate the feeding systemic arterial vessel(s), nor check up for it during surgery.

The interesting part of our case was that it proposed that the elevated serum CA 19-9 was very likely related to the prenatal detected retroperitoneal pulmonary sequestration. CA 19-9 concentrations can be increased in patients with various lung abnormalities [5]. We located adult RPS case [6] and PS cases with elevated CA 19-9 serum levels, and return of the elevated CA 19-9 values to normal after lesion removal. In the adult RPS case, immunohistochemical study demonstrated CA 19-9 in the bronchial mucosa of the lesion. There is no report about the relationship between CA 19-9 and prenatal detected RPS. An early histological and immunohistochemical analysis of lung development showed cells positive for CA 19-9 appeared during the time when bronchial branches developed, and were abundant at the time of completion of bronchial branching [7]. It was reasonable that the dysplastic lung tissue in our case cause abnormal CA 19-9 serum level. While there was no report about the abnormal serum CA 19-9 level in infants with other lesion in the area, we hope this finding can help with the differential diagnosis in some other cases.

It is important to get prenatal detected RPS distinguished from more common fetal and neonatal lesions in adrenal region: neuroblastoma and adrenal hemorrhage, because they do not share same treatment strategy. With review of the literature, we found that an algorithm posed by Pablo Laje [3] is practical, but it doesnt cover every case. Its designer(s) seemed not to know the fact that in the prenatally detected population, sensitivity of the urinary catecholamines assay for neuroblastoma was only 36%. It is disturbing that the urinary catecholamines assay for neuroblastoma can be false positive in RPS [8]. The sonographic appearance of prenatal adrenal hemorrhage does not always follow a typical course. The detection of the feeding vessels for RPS could fail due to not-advanced Imaging modalities or when it was fed by small branches instead of dominant vascular stalk [9]. We may have to look for the histological evidence.

Fig. 1. A: The mass was hyperechoic, measured 2.3 × 1.5 cm at 28 weeks’ gestation. It was superior, medial to the left kidney, below the diaphragm. B: A magnetic resonance imaging (MRI) scan at the same time showed the lesion was hyperintense on T2-weighted. C: Computerized tomography (CT), at 6 weeks after birth, showed a non-calcified heterogeneously enhanced solid mass in the left adrenal gland region. D: The histologic study of the specimen revealed the presence of alveolar-like squamous cells (S). Simple ciliated columnar epithelium (C), and smooth muscle fiber (arrow), which was consistent with dysplastic lung tissue. E: Positive immunostaining with an anti-CA 19-9 antibody.
The treatment remains controversial, mainly because the prenatal detected RPS has potential to resolve spontaneously [10], as well as to malignant transformation especially when coexists with congenital cystic adenomatoid malformation (CCAM) [11]. Most RPS are asymptomatic, although they link to potential complications, like compression of the normal structures [4] causing dysphagia, GERD, and RDS, pheochromocytoma-like symptoms [12], or high flow heart failure caused by the systemic blood supply [4]. We agree with the surgical indication of symptomatic lesions. If it is not, because of the possibility of its spontaneous postnatal involution, observation may be proper for a while to avoid surgery. It may at least require 9 month until the lesion start to regress [10]. Since CCAM, with which ELS is frequently associated [13], may experience the malignant transformation, and the transformation can be found as early as 22 months after born [9], it is worrying to apply the strategy of “wait and see” so long. Surgery removal is also required to avoid some future problems like infection [4], abdominal pain [14]. When surgery is decided, laparoscopic approach is better for the sake of minimal invasive.

3. Conclusion

RPS is a rare disease. It is important to be distinguished from neuroblastoma for RPS, although sometimes it can be difficult. Our finding that the RPS in some cases may lead to elevated serum 19-9 level can help with the differential diagnosis. Strategy of “wait and see” should be applied firstly when there is no related symptom. Surgery is only needed when spontaneous regression does not take place.

Conflict of interest

The authors declare no conflict of interest.

References