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Brief Report

Left atrial extension of hepatoblastoma via left superior pulmonary vein

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Abstract Hepatoblastoma is the most common malignant liver tumour in early childhood. The metastatic extension of hepatoblastoma into the left atrium via the pulmonary vein is rare. Reported lesions almost always involve a right-sided approach. Here we report the case of a 3-year-old girl with a recurrent hepatoblastoma at multiple sites, including the left atrium, brain, and lung. The patient was treated surgically for the prevention of further embolic complications and cardiac failure.

Keywords: Hepatoblastoma; paediatric heart surgery; left atrial mass; cardiac failure

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Benign cardiac neoplasms occur three times more than malignant tumours. Cardiac tumours were first described in the 18th century by Boneti, and in 1936 the first successful removal of a neoplasm of the heart was performed.[1] Malignant tumours that originate elsewhere in the body and metastasize to the heart are more common than ones that originate in the heart. Left atrial and left ventricular tumours can present with various signs and symptoms. These tumours may embolise and they can also lead to seizures, transient ischaemic attacks, and cerebrovascular and peripheral-vascular accidents. On the basis of their size and position, they may induce arrhythmias and interfere with ventricular compliance.

Hepatoblastoma is a hepatic tumour predominantly occurring in children. The usual site of metastasis is the lung. Metastasis of hepatoblastoma to the brain and the left side of the heart are quite rare. Here we present a rare case of a hepatoblastoma involving the pulmonary vein with extension into the left atrium and brain metastasis.

Case report

A 3-year-old girl was admitted with a 4–5-week history of headache, irritability, and decreased appetite with a 4–5-day history of vomiting. She had a previous history of hepatoblastoma diagnosed 2 years earlier, which was confirmed by a computed tomography-guided biopsy. At that time, four cycles of pre-operative chemotherapy consisting of cisplatin, 5-fluorouracil, and vincristine were administered and tumour resection was done. The patient apparently remained stable for the following 1 year until her present symptoms developed. However, in the intervening period, she did not undergo follow-up in spite of the physician's advice.

Owing to persistence of her symptoms, chest computed tomography and brain magnetic resonance imaging were performed. The patient had an abnormal mass-like lesion occupying the left upper lung field on a plain chest X-ray. Her breath sounds were markedly decreased in the left upper chest. There was no palpable abdominal mass. Brain magnetic resonance imaging revealed a temporal haemorrhagic brain metastasis with extensive peri-lesional parenchymal oedema. Chest computed tomography showed a heterogeneous mass measuring 4.5×4 cm in the left superior lobe and 6×3 cm in the right inferior lobe of the lung. Computed tomography also revealed a hypodense mass measuring 3.5 cm, which was consistent with a direct invasion into the left atrium or a thrombus-like lesion involving the left atrium. At this point, the patient's serum alpha-feto protein level was 24,363 ng/ml. All these findings confirmed the diagnosis of recurrent hepatoblastoma

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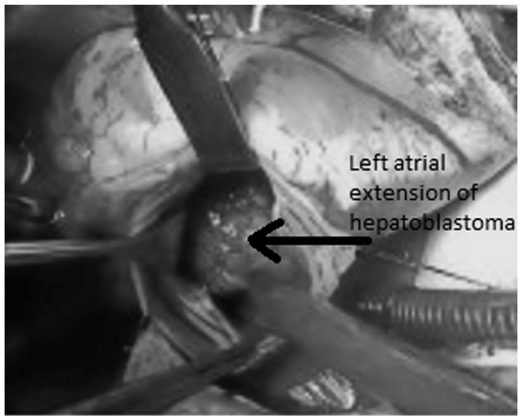

Figure 1.
Image of the metastatic mass of hepatoblastoma.

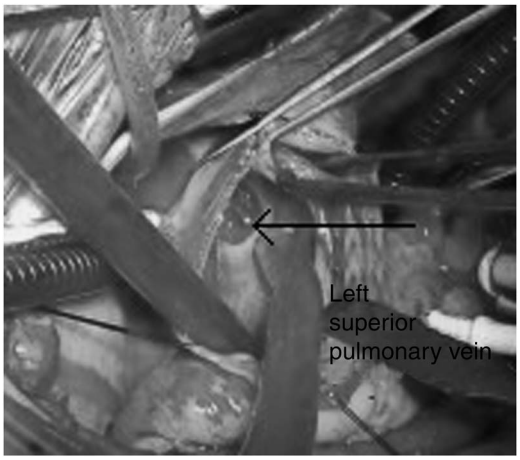

Figure 2.
Peri-operative image of the large pedunculated, fibrous, hard mass.

with brain and lung metastasis. A needle aspiration of the pulmonary mass revealed histology consistent with the diagnosis of hepatoblastoma. An echocardiogram confirmed two large masses in the left atrium, measuring 3×3 cm and 1×1 cm, causing mitral valve obstruction and 2\4 mitral insufficiency. Both masses appeared to originate from the left superior pulmonary vein. The remaining cardiac structures were normal on echocardiography. Palliative surgical intervention was undertaken to relieve the pulmonary venous obstruction caused by the tumour extension. Median sternotomy, aortic-bicaval cannulation, and standard cardiopulmonary bypass were performed for excision of the left atrial mass. For surgical removal of the left atrial mass, we used the extended vertical transatrial septal approach. The large pedunculated, fibrous, hard mass with smooth surface occupying one-third of the left atrium was excised; the pedicle was traced to the left superior pulmonary vein (Fig 1). The mass was adherent to the atrial wall by fibrous tissue, which was resected. It extended from the left superior pulmonary vein, totally occluding the lumen (Fig 2). Similar tissue extending into the inferior right pulmonary vein was excised. The inferior left pulmonary vein appeared normal. The early post-operative recovery was uneventful; the patient reported feeling well with no evidence of tumour emboli or myocardial dysfunction. She had – vincristine, 5 FU, and cisplatin – adjuvant chemotherapy and radiotherapy 10 days after the operation. Death occurred 7 months later because of severe sepsis and septic shock.

Discussion

Tumour infiltration of the great vessels is not uncommon, and a number of instances of primary pulmonary malignancies with intra-left atrial extension via the pulmonary vein have been reported. Takahashi et al[2] found 20 patients with tumours potentially invading the proximal portion of the pulmonary veins and left atrium. In contrast to the above findings with primary tumours, however, left atrial extension of metastatic lung tumours through pulmonary veins is extremely rare. The most common mechanism of intracardiac involvement from hepatoblastoma involves direct extension of the tumour via the inferior vena cava into the right atrium, and possible extension to the patent foramen ovale and the left atrium. It is also possible that left atrial metastasis could occur by haematogenous spread of tumour and the tumour cells spread from the tumour located in the lung via pulmonary veins directly into the left atrium.

The most frequent site of metastasis of hepatoblastoma is the lung. Lung metastases are the most reliable prognostic factor of an unfavourable outcome for patients with hepatoblastoma. Extra-pulmonary metastasis of hepatoblastoma, including brain and heart, is uncommon. However, cardiac metastases, which most often involve the right side of the heart, are uncommon and difficult to diagnose and are most often found at autopsy. However, there were multiple brain lesions and involvement of the left side of the heart in our case.

Collins et al[3] reported that widespread embolism occurred soon after detection of the intracardiac extension of leiomyosarcoma. In another case, an intra-atrial lesion induced syncope and palpitations.[4] Given the severe complications that have been reported in patients with this type of extension, as in our case, immediate prophylactic surgery should be considered.

We did not resect the lung lesion because of the criteria for resection of metastatic lung lesions. There are five primary criteria for surgical resection of pulmonary metastases: (1) no disease at the site of the

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primary tumour; (2) no metastases outside the lungs; (3) no non-resectable pulmonary nodules; (4) no non-surgical alternative for cure; and (5) no evidence that the patient cannot tolerate the proposed surgical therapy.[5] In the present case, the patient had multiple unresectable metastases. We resected the heart mass because of circulatory considerations, not for cancer cure.

In our case, we felt that the growing tumour might induce sudden cardiac arrest owing to obstruction of cardiac inflow and that resection should also be performed to prevent embolic complications. We thus recommended immediate prophylactic surgical resection of the tumour.

Conclusion

We report a case of metastatic lung tumour extending into the left atrium via the pulmonary vein. Hepatoblastoma exhibiting this type of extension is extremely rare. Given the severe complications that have been reported in patients with this type of tumour extension, immediate prophylactic surgery should be considered.

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