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## CASE REPORT

# Congenital hepatoblastoma in a growing health economy

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**SUMMARY**

A 43-year-old woman, whose pregnancy was complicated by the presence of a large single solid intra-abdominal fetal mass, was referred from the private sector into our fetal maternal unit at the Corniche Hospital, Abu Dhabi at 36 weeks postmenstrual age. Investigations subsequently confirmed that this mass was a congenital hepatoblastoma, one of the very rare embryonic tumours. The baby had chemotherapy and surgical excision of the tumour. Fifteen months later, the alpha fetoprotein levels remain normal and follow-up MRI scans do not show recurrence or any residual disease.

To our knowledge, this is the first case of congenital hepatoblastoma in the United Arab Emirates (UAE). In the UAE, the interphase between private health insurance schemes and medical (public and private) care within a growing health economy enhances access to unique services such as cancer treatments within specialised centres.

**BACKGROUND**

We believe this case is important because it's the first case of congenital hepatoblastoma to be reported from the United Arab Emirates (UAE). The management reflects the changing dynamics in the healthcare infrastructure in the Emirates with availability of highly specialised treatments within the country guided by international protocols and thereby reducing the dependence on external health systems.

**CASE PRESENTATION**

A 43-year-old, Gravida 6 Para 3, woman was referred to our fetal-maternal team at 36 weeks of gestation because the fetus was noted to have an unusual large intra-abdominal mass. She had diet controlled gestational diabetes and rheumatoid arthritis controlled with hydroxyquinone. Parents are non-consanguineous.

The antenatal ultrasound scan showed the presence of a huge echogenic abdominal mass (7.8×7.2×7.7 cm). The abdominal circumference was 35.7 cm (>97.7 centile for 36+4 weeks postmenstrual age). There was no evidence of significant arteriovenous shunting. Fetal arterial and venous Doppler's and fetal echocardiography were normal. There was no evidence of hydrops fetalis. Pulmonary fields, brain and placenta appeared sonographically normal. Delivery at 39 weeks was

planned as there was no impact of this mass on fetal well-being.

A female child weighing 3440 g (50th centile) with occipital-frontal circumference of 34 cm (50th centile) was born following elective caesarean section at 39 weeks. Apgar's were 9 and 10 in 1 and 5 min, respectively. The presence of a large abdominal mass was evident immediately after birth. She did not require any respiratory support but had bile stained gastric aspirate/vomiting due to a mass pressure effect on the bowel which resulted in delays in establishing enteral feeds. CT of abdomen and pelvis was obtained within a few days after birth which identified a highly vascular mass measuring 8.6×7.1×7.8 cm that appeared to be of hepatic origin.

**DIFFERENTIAL DIAGNOSIS**

Neuroblastoma, nephroblastoma, vascular tumours and rhabdoid tumours were considered as possible differentials antenatally. It was difficult to ascertain and assign the origin on antenatal ultrasound due to the gestational age at which mother presented to our fetal maternal unit.

**INVESTIGATIONS**

- ▶ Alpha fetoprotein (AFP) measured over 68 000 IU/L on day 8 of life. It was serially monitored during the treatments. Normal AFP level would be expected to be between 20 and 30 IU/L within a few weeks after birth.
- ▶ On day 12 of life, laparoscopic tumour biopsy was obtained which was positive for Hep-par 1 and AFP. These results (received a week later) confirmed that the mass was a hepatoblastoma (epithelial type with a fetal pattern).
- ▶ Pre-chemotherapy CT of the chest revealed no evidence of metastasis. The tumour was a PRETEXT II (V and P negative).<sup>1</sup>

**TREATMENT**

Hepatoblastoma was confirmed on day 19 of life. Chemotherapy was commenced on day 24 of life after baseline investigations which included cardiac echocardiography and audiology screen were completed.

The Children's Oncology Group Protocol called AHEP0731 was applied. This consists of cisplatin, 5-fluorouracil, vincristine and doxorubicin delivered every 21 days over six cycles. The tumour was excised at 14 weeks postnatal age after the second cycle of chemotherapy.



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Following the second course of chemotherapy, CT showed a reduction of the tumour size to 6.4×6.2×5.5 cm and a corresponding fall in the AFP levels from just over 68 000–2900 IU/L. The tumour was staged as POST-TEXT II (V and P negative) at this time point.

Complete surgical excision of the tumour was undertaken at 14 weeks' postnatal age following which the AFP levels fell further to 303 IU/L.

### OUTCOME AND FOLLOW-UP

She was discharged home after the third course of chemotherapy and then electively readmitted for subsequent courses.

Post chemotherapy morbidities that occurred after the first cycle included profound neutropenia (associated with fever on two occasions). She also developed vincristine-induced polyneuropathy involving the median nerves, peroneal nerve and sensory nerves of the upper limbs. She lost gross motor skills and developed an unsafe swallow. Vincristine was thereafter given at 50% of the dose in subsequent courses of chemotherapy. The neuropathy gradually resolved allowing the safe resumption of oral feeding 3 months after chemotherapy had been initiated.

There was no evidence of disease on serial MRI scans of the liver following the surgical resection. At the age of 19 months (12 months after the last course of chemotherapy) the AFP remains normal. Follow-up, which will include regular monitoring of AFP levels, will be for at least the next 5 years.

### DISCUSSION

The UAE is a young country that is growing and developing its infrastructure in several domains including Paediatric Oncology. This is supported by investments in fetal–maternal expertise, neonatal intensive care and cancer treatments across the Emirates. The case described is, to our knowledge, the first reported case of congenital hepatoblastoma in the UAE.

Hepatoblastoma is the the most common primary paediatric hepatic malignant tumour (although comparatively, they are one of the very rare paediatric solid tumours) with a reported frequency of 1–11.2 per million children under the age of 15 years.<sup>2</sup> Most commonly, they arise from the right lobe of the liver.<sup>3</sup> They may be associated Beckwith-Wiedemann syndrome and familial adenomatous polyposis.<sup>3</sup>

Antenatal diagnosis of intra-abdominal tumours and hepatoblastoma has been reported previously.<sup>4</sup> However, antenatal ultrasound diagnosis of congenital abdominal tumours is difficult and assigning their origin, especially at late gestational ages, is very challenging. Comprehensive Doppler studies and additional three-dimensional ultrasound techniques may help in this task, in order to prepare and optimise delivery. Fetal MRI in the third trimester may help to determine size and relation of the tumour to neighbouring organs, if specialised image interpretation is available.<sup>4</sup> In general, the presence of fetal abdominal tumour does not affect time and mode of delivery; standard obstetric antenatal and intra-partum care are warranted. However, cases need to be individualised, since excessive tumour growth may lead to significantly increased abdominal circumference that could, in a vaginal delivery, increase the risk of tumour rupture and increase the chances of dystocia.<sup>4</sup> Elective caesarean section may be preferred for tumours diagnosed in the antenatal period as suggested by Trobaugh-Lotrario *et al*<sup>5</sup> following their review of a large series of their cases.<sup>5</sup>

Within the UAE, Standardised Cancer treatment protocols are used for treating children with cancer<sup>6</sup> within a highly specialised

### Patient's perspective

"Very happy especially, that as not from this country, my baby have treatment and is now good. Thank you"

### Learning points

- ▶ First reported case of congenital hepatoblastoma from the United Arab Emirates to our knowledge.
- ▶ The importance of multidisciplinary teams—fetal maternal, neonatology, paediatric surgery, oncology and parents working in partnership to deliver care.
- ▶ The interphase between health insurance schemes and private and public-sector healthcare should be seamless—allowing all individuals access to specialised services.
- ▶ The evolving health infrastructure in the Middle East allows access to specialised treatments within the county therefore reducing physical dependence on external systems.

multi-disciplinary setting. Treatment of hepatoblastoma depends on the staging which consists of chemotherapy and surgery in the stable patient.

This report highlights the importance and role of an evolving interphase between private and public healthcare and private insurance schemes. This relationship facilitates and promotes multidisciplinary team-working in the UAE, allows the application of expertise to make the diagnosis of such rare conditions and gives access to appropriate treatments in order to achieve good clinical outcomes.

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