Pediatric Radiology Case Challenge: An Educational Activity for AOSPR 2022 Attendees

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ABSTRACT

The following cases presented in this article were originally part of an interactive online quiz for the participants of the 20th Asian and Oceanic Society for Paediatric Radiology (AOSPR) Annual Scientific Meeting held virtually on August 19 and 20, 2022. The objectives for the quiz were to direct self-education of the attendees, to introduce confounding cases and test diagnostic ability, and to review and discuss the pertinent findings of the selected cases.

Included are seven cases showing various cranial and abdominal computed tomography (CT) and magnetic resonance imaging (MRI) studies, which range from mundane to the complex. Each case is followed by a brief discussion of relevant findings and differential diagnoses.

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CASE 1

A 12-day-old boy delivered term by cesarean section after prolonged labor was referred for a brain CT scan due to an enlarging head circumference.

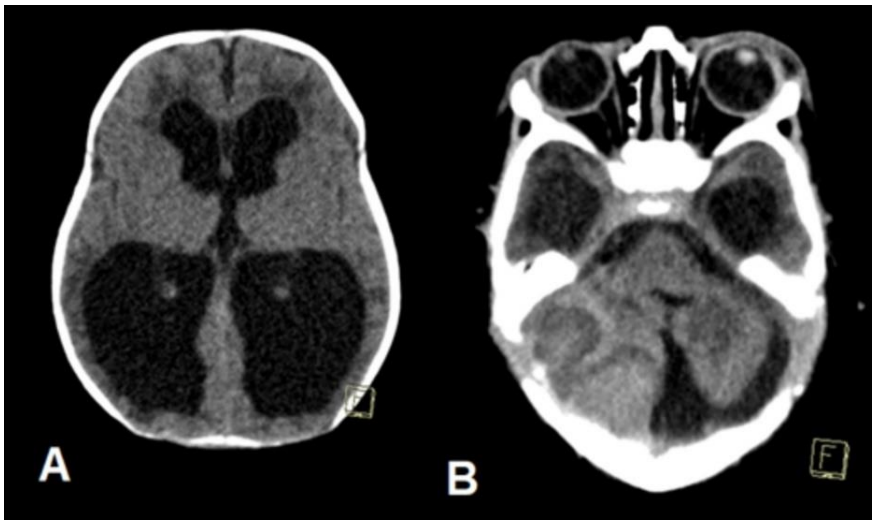

Fig. 1A–B Non-contrast axial CT images of the brain show (A) hydrocephalus, and (B) heterogeneous extra-axial collection compressing the adjacent right cerebellum

Question: Which of the following is NOT true regarding the etiology of the imaging findings?

A. More common in term infants (correct answer)
B. Ultrasound could be used to make an early diagnosis
C. MRI can identify the anatomic structures better than CT
D. None of the above

Posterior fossa abnormalities are common in high-risk term infants, seen in about 66%, the most common of which is hypoxic-ischemic encephalopathy. Central nervous system (CNS) malformation, infection and posterior fossa hemorrhages are also commonly identified [1].

In the case presented, the collection at the right posterior fossa (Fig. 1B) shows mass effect on the cerebellum, for which mass lesion and hemorrhage are initially considered. The neonate was clinically suspected to have a bleeding disorder such as hemophilia and referred for genetic testing, which made posterior fossa hemorrhage the most likely diagnosis. Posterior fossa hemorrhage in term neonates is rare and more common in preterms, however incidence may range from 3.4% to 4% in hemophiliacs [2]. It is also difficult to detect since clinical symptoms are non-specific and may include increased head circumference, seizures or respiratory distress. Respiratory symptoms may occur when the medulla oblongata is involved. In neonates suspected with hemorrhage, cranial ultrasound would usually be the initial imaging examination of choice.

CASE 2

A 4-month-old boy was admitted for pneumonia. The infant was also assessed to have developmental delay; hence imaging was performed.

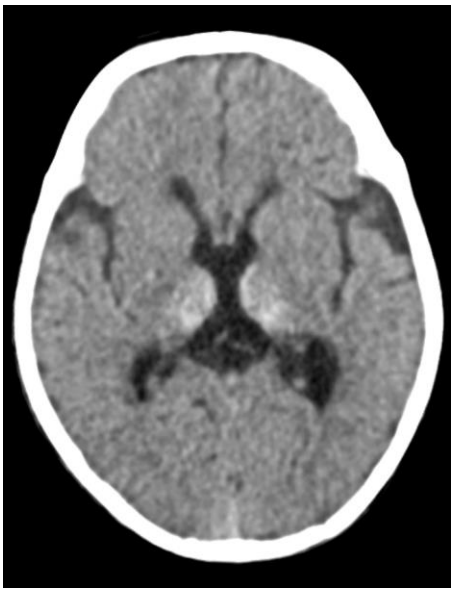

Fig. 2 Non-contrast axial CT image of the brain shows an unusual hyperdensity of the thalami

Question: Hyperdensity of the thalami can be seen in?

A. Profound perinatal ischemia
B. Krabbe disease
C. Gangliosidosis
D. Glioma
E. All of the above (correct answer)

All of the choices would result in the finding of thalamic hyperdensities (Fig. 2) in this infant. Differential diagnoses would include inherited metabolic disorders (Lysosomal storage disorders/gangliosidoses, Krabbe disease), acquired toxic/metabolic, infectious, neoplastic (glioma, lymphoma), or hypoxic-ischemic encephalopathy. Perinatal ischemia should be suspected since the deep brain structures, including the thalami and basal ganglia [3] are the most metabolically active and are therefore most vulnerable, especially in the first 3 days of life. Ischemia secondary to the patient's pneumonia is less considered since CT hyperdensity is considered a late finding, while acute hypoxic-ischemic changes should present as hypodensity. CT scan however is less sensitive and ideally MRI should be performed.

Lysosomal storage disorders such as Krabbe disease would present as bilateral thalamic CT hyperdensity [4] early on, as well as the caudate nuclei, corona radiata and the cerebellar dentate nuclei. MRI with diffusion tensor imaging and spectroscopy may also be useful for early diagnosis and to detect the extent of white matter involvement. Imaging findings may however overlap with other leukodystrophies and hypoxic-ischemic encephalopathy.

CASE 3

An 8-year-old boy who was previously diagnosed with neurofibromatosis type I (NF-1) underwent an MRI study due to a 6-week history of headache and blurring of vision.

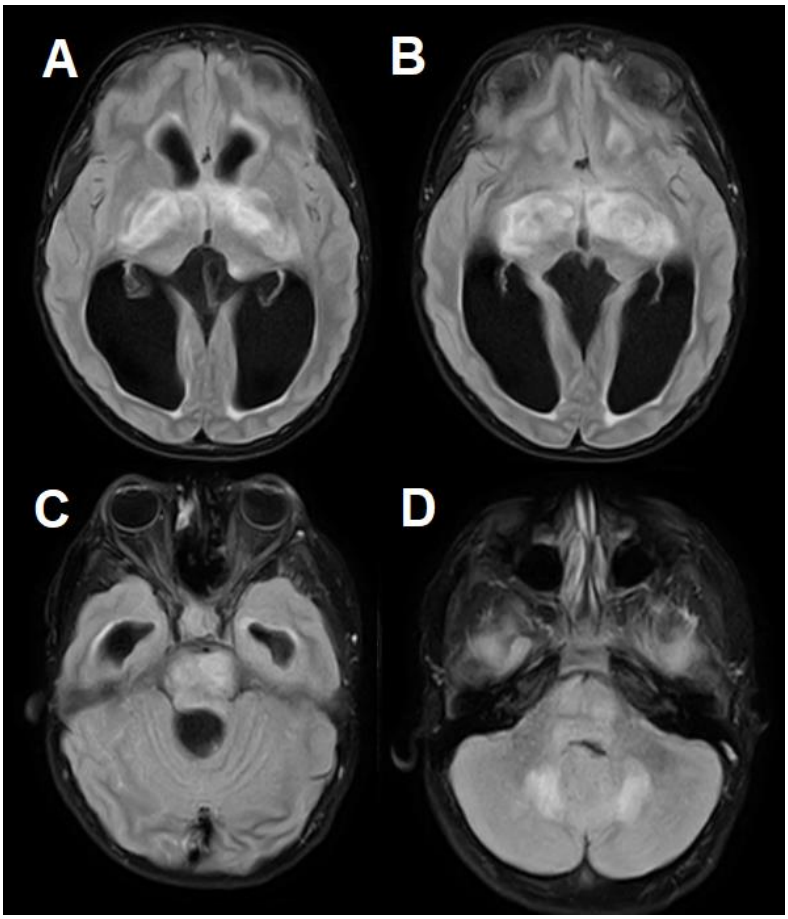

Fig. 3A–D Non-contrast axial MR images of the brain in fluid-attenuated inversion recovery (FLAIR) sequence show hyperintensities at the (A,B) bilateral globus pallidi, (C) midbrain, pons, and (D) dentate nuclei. Effaced fourth ventricle with hydrocephalus and transpendymal edema also detected

Question: What is the most commonly seen CNS MRI finding in Neurofibromatosis type I?

The most common brain lesions found in NF-1 on MRI are abnormal T2W hyperintensities, which are coined as focal areas of (high) signal intensity (FASI), without or with minimal mass effect. These rarely enhance after gadolinium infusion. These can be found in about 60% to 70% [5] or 60% to 80% [6] of pediatric NF-1 patients. MRI is the imaging modality of choice for detection of FASIs since these lesions are not usually seen on CT scan. The main differential diagnosis would be low-grade glioma which can present with more evident mass effect and enhancement.

The clinical significance of FASIs is largely unknown, and are believed to be caused by increased fluid accumulation in intramyelinic vacuoles. These lesions tend to decrease during adolescence, for which the reason remains unclear. The presence of concomitant optic gliomas within and outside the optic pathways may also muddle interpretation.

CASE 4

A 2-year-old girl presented with 4 days of abdominal pain and vomiting, and was admitted as a case of acute gastroenteritis. CT scan was done showing ileocolic intussusception.

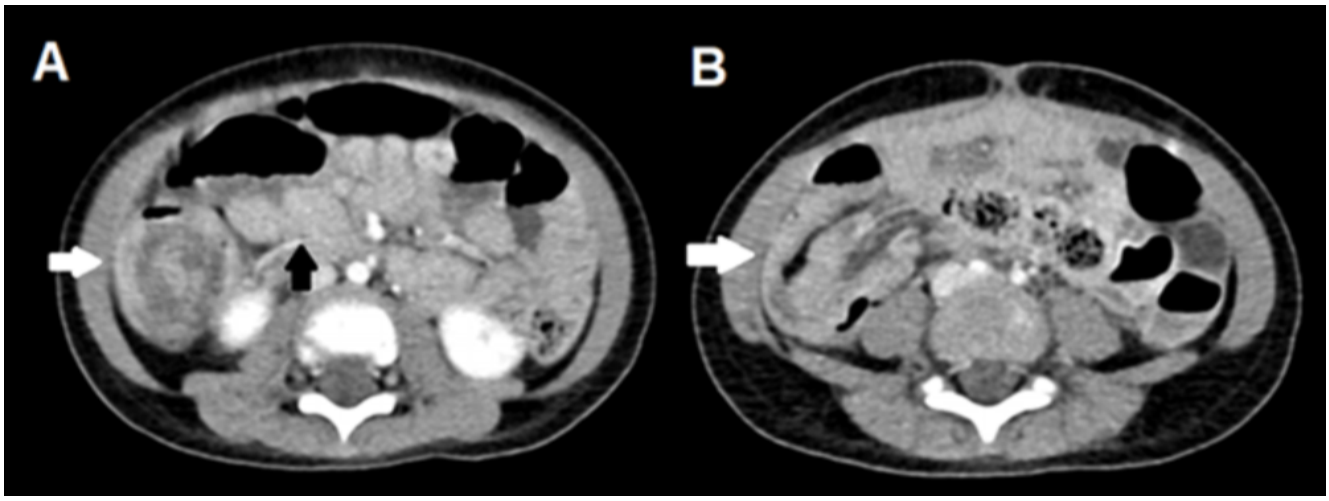

Fig. 4A–B Contrast-enhanced axial CT scan demonstrates (A) mesenteric adenopathy at the right hemiabdomen, with a 'target' lesion (A, B, white arrows) demonstrating ileocolic intussusception. There is (B) telescoping of the ileocecal region into the intussuscipiens

Question: The most likely lead point for intussusception is:

A. Lymphoid hypertrophy
B. Mesenteric lymph node
C. Juvenile Polyp
D. All of the above (correct answer)

Ileocolic intussusception is the most common type, seen in 75% to 95% of cases, with most of the causes being idiopathic. 1.5% to 12% [7] present with a pathologic lead point, while 90% of cases present with a lead point in adults. 66% of lead points are detectable during ultrasonography. The most common pathologic lead point is from Meckel diverticulum. The presence of a Meckel diverticulum is not a contraindication to air enema but there is an increased risk of recurrence of about 78.5%. Contraindications to fluoroscopic or ultrasound-guided reduction are the presence of pneumoperitoneum or peritonitis.

CASE 5

This is a case of a 12-year-old girl with 3-month history of abdominal bloatedness and discomfort.

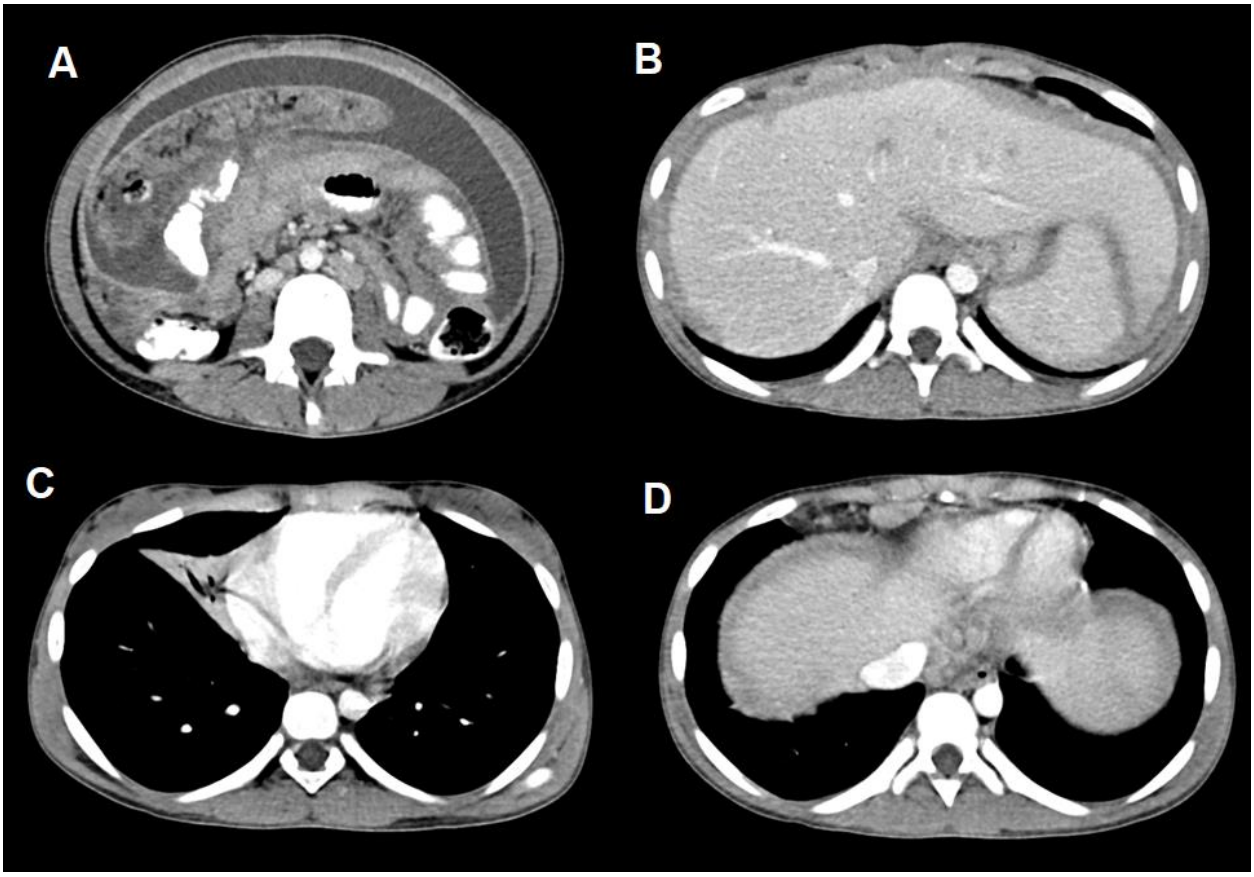

Fig. 5A–D Contrast-enhanced axial CT images demonstrate (A) ascites with smooth peritoneal thickening, as well and nodular mesentery. Other findings include (B) multifocal hepatic hypodense nodules, (C) incidental note of middle lobe atelectasis and (D) cardiophrenic lymphadenopathy

Question: What is the diagnosis based on the presented CT images?

A. Peritoneal carcinomatosis
B. Primary peritoneal mesothelioma
C. Peritoneal tuberculosis (correct answer)

Malignancy cannot be entirely ruled-out when faced with multi-organ involvement on imaging, particularly when presented with non-specific symptoms. Histopathologic findings in this patient would however prove the presence of extra-pulmonary tuberculosis. Abdominal involvement occurs in 11% to 12% of patients with extra-pulmonary tuberculosis [8], while peritoneal tuberculosis is the most common presentation. Based on the CT findings, a wet type of tuberculosis is considered, due to presence of ascites with peritoneal and mesenteric thickening.

Peritoneal tuberculosis would present as ascites, as seen in 70% to 90% [8], followed by peritonitis (38%), lymph node disease (23%), gastrointestinal (19%) and solid organ (10%) involvement. The most common solid organs involved are the liver and spleen in 70%, such as in our patient with hepatic nodules. The presence of mesenteric nodularity and peritoneal thickening in this case proved to be a diagnostic dilemna, since this mimics peritoneal carcinomatosis. With this in mind, the differential diagnoses of lymphoma and adenocarcinoma are not entirely ruled-out even in the pediatric population.

CASE 6

A 17-year-old boy with persistent abdominal pain despite endoscopic retrograde cholangiopancreatography (ERCP) for a known choledocholithiases.

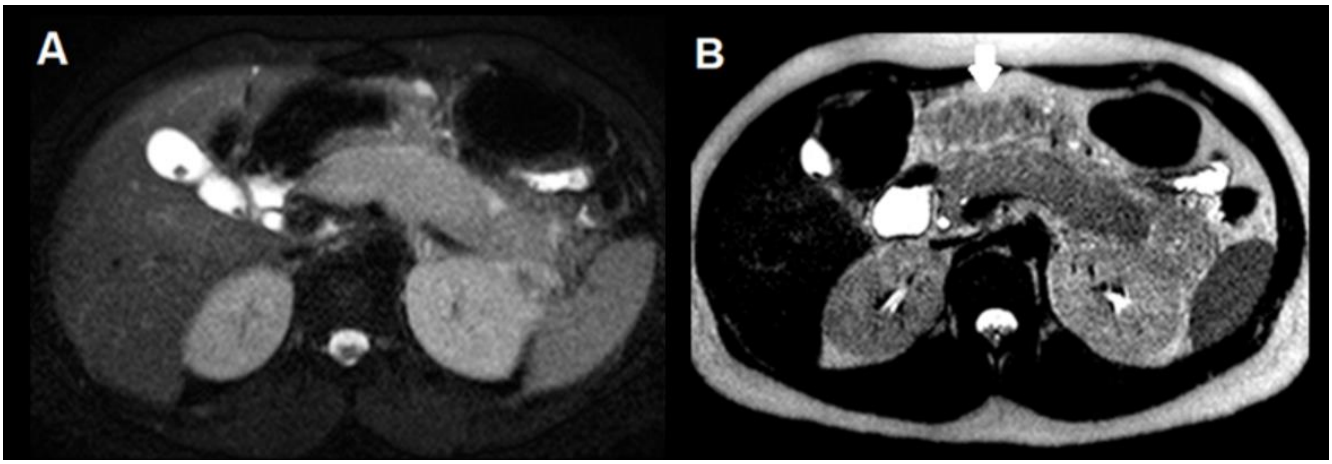

Fig. 6A–B Axial MRI of the abdomen reveals (A) cholelithiases and edematous pancreas with minimal peripancreatic fluid, while (B) there are also T2-weighted isointense and nodular peripancreatic and omental fat planes (white arrow)

Question: What would account for the unusual appearance of intra-abdominal fat?

A. Peritoneal carcinomatosis
B. Liposarcoma
C. Fat necrosis (correct answer)
D. Retroperitoneal fibrosis

The patient on MRI had findings suggestive for pancreatitis, therefore fat necrosis should be considered since this is a known complication of acute pancreatitis [9]. This is associated with low (12% mortality), and may develop over weeks to several months. The typical distribution of fat necrosis is peripancreatic, and extends to the fat planes of the transverse mesocolon, omentum and the mesenteric root. Fat necrosis is secondary to ghost fat cells damaged by pancreatic enzymes. Release of fatty acids precipitate with serum calcium, producing the phenomenon of saponification [10] of intra-abdominal fat.

Eventually when the acute phase of pancreatitis subsides, fat necrosis / saponification would take on the appearance of nodular lesions throughout the retroperitoneum and mesentery. Adding to confusion of those unfamiliar with the patient's history, imaging findings may mimic carcinomatosis while encapsulated fat necrosis can mimic liposarcoma.

CASE 7

A 15-day-old girl was known to have a hepatic mass seen during a congenital anomaly scan, likely a hemangioma. The baby was born with APGAR score of 6 and 8, and diagnosed with patent ductus arteriosus and congestive heart failure.

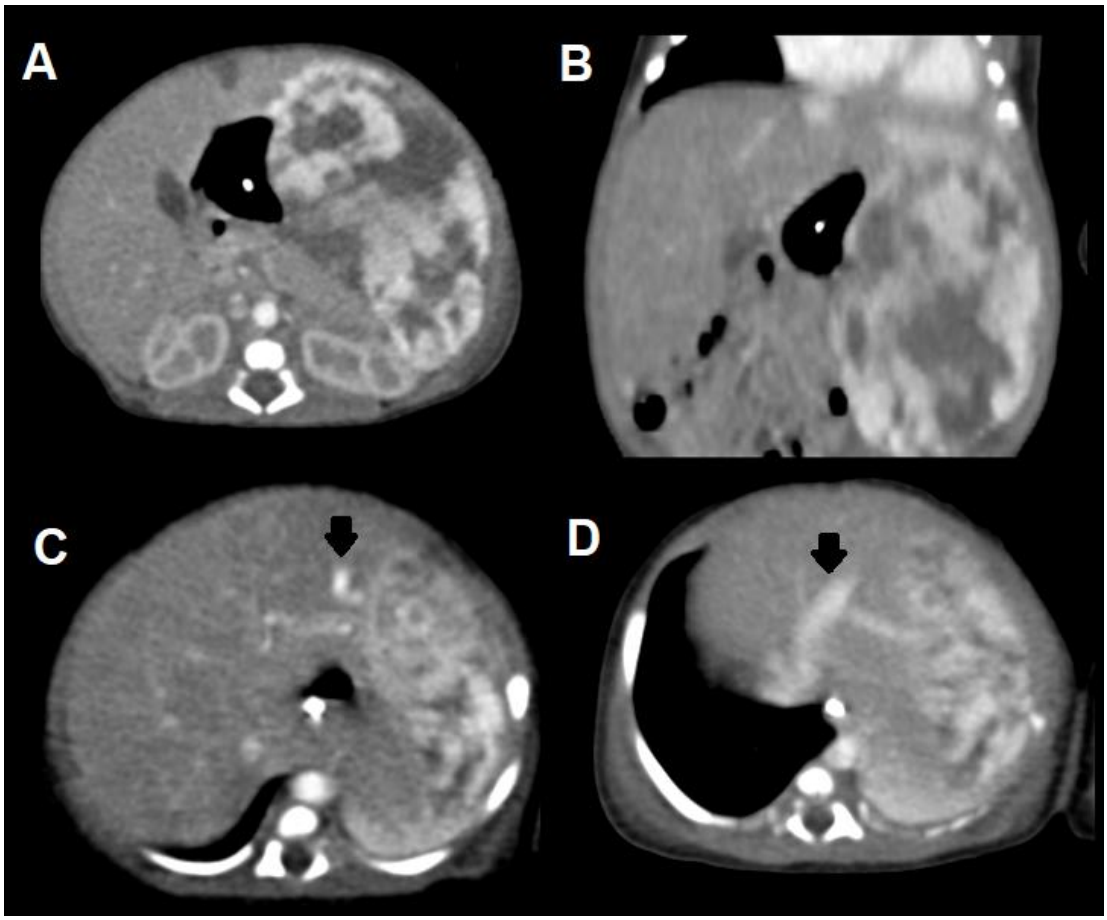

Fig. 7A–D Contrast-enhanced axial CT images of the abdomen show a (A,B) large vascular mass occupying the left hepatic lobe. (C) Arterial feeding vessels from the left hepatic artery (black arow), and (D) dilated draining left hepatic vein close to the lesion are identified, reflective of arteriovenous shunting

Question: Which imaging finding seen in this patient with Infantile Hepatic Hemangioma contributes to congestive heart failure?

Arteriovenous shunt (correct answer); An infantile hepatic hemangioma (IHH) is commonly asymptomatic, but 15% [11] of cases present with high-output cardiac failure with mortality of 70% to 90%. In the presented CT scan images, an arteriovenous shunt is present, which is the pathologic mechanism of congestive heart failure for patients with IHH. The shunt results in decreased systemic blood volume, with subsequent increased pulmonary blood volume (pulmonary hypertension) and resulting in cardiac output increase. 70% of all IHH-related deaths are caused by congenital heart failure [12], hence it is imperative that the arteriovenous shunts are reduced.

Treatment may be medical such as giving oxygen support, sedatives, diuretics and steroids. Transcatheter arterial embolism (TAE) is also strongly suggested for stabilization of heart failure in conjunction with pharmacologic therapy. For large hemangiomas and shunts however, these remain to be stop-gap measures with definite treatment being surgical resection and liver transplantation in some cases [11].

REFERENCES

1) Steggerda SJ, De Bruïne FT, Smits-Wintjens VE, Verbon P, Walther FJ, Van Wezel-Meijler G. Posterior fossa abnormalities in high-risk term infants: comparison of ultrasound and MRI. Eur Radiol 2015;*25*(9):2575–2583. https://doi.org/10.1007/s00330-015-3665-8.
2) Tsai P, Chen H, Ho C, Chiu N. Posterior fossa hemorrhage in a term neonate with hemophilia A. J Med Ultrasound 2018;26(1):56–58. https://doi.org/10.4103/JMU.JMU_10_18.
3) Heinz ER, Provenzale JM. Imaging Findings in Neonatal Hypoxia: A Practical Review. AJR Am J Roentgenol 2009;192(1):41–47. https://doi.org/10.2214/ajr.08.1321,
4) Van Cauter S, Severino M, Ammendola R, et al.. Bilateral lesions of the basal ganglia and thalami (central grey matter) — pictorial review. Neuroradiology 2020;62(12):1565–1605. https://doi.org/10.1007/s00234-020-02511-y.
5) Calvez S, Levy R, Calvez R, et al. Focal Areas of High Signal Intensity in Children with Neurofibromatosis Type 1: Expected Evolution on MRI. AJR Am J Neuroradiol 2020;41(9):1733–1739. https://doi.org/10.3174/ajnr.a6740.
6) Petrak B, Lisy J, Kraus J, Kyncl M, Zatrapa T. Focal areas of high-signal intensity on brain T2-weighted magnetic resonance imaging scans are significant for the diagnosis of neurofibromatosis Von Recklinghausen type 1. Pediatrics 2008;121(Suppl 2):S147. https://doi.org/10.1542/peds.2007-2022cccccc.
7) Dennis R, Anupindi S, Khwaja A. How to Diagnose Intussusception in Children. Appl Radiol 202150(1);19–27. https://doi.org/10.37549/ar2699.
8) Da Rocha EL, Pedrassa BC, Bormann RL, Kierszenbaum M, Torres L, D'Ippolito G. Abdominal tuberculosis: a radiological review with emphasis on computed tomography and magnetic resonance imaging findings. Radiologia Brasileira 2015;48(3):181–191. https://doi.org/10.1590/0100-3984.2013.1801.
9) Smith JR, Arnoletti JP, Varadarajulu S, Morgan DE. Post-pancreatitis Fat Necrosis Mimicking Carcinomatosis. Radiol Case Rep 2008;*3*(2):192. https://doi.org/10.2484/rcr.v3i2.192.
10) Devos H, Goethals L, Belsack D, et al. Fat misbehaving in the abdominal cavity: a pictorial essay. Pol J Radiol 2020;85(1):32–38. https://doi.org/10.5114/pjr.2020.93070.
11) Zhang X, Ren W, Song G, Xiao Y, Sun F, Wang N. Infantile hepatic hemangiomas associated with high-output cardiac failure and pulmonary hypertension. BMC Cardiovasc Disord 2019;19(1):216. https://doi.org/10.1186/s12872-019-1200-6.
12) Wang T, Wang Y, Liang Y, Lu G. Infantile Hepatic Hemangioendothelioma Associated with Congestive Heart Failure. Medicine (Baltimore) 2015;94(52):e2344. https://doi.org/10.1097/md.0000000000002344.