Duodenal Web with Trichobezoar: An Unusual Presentation

Duodenal atresia and duodenal web cause upper gastrointestinal obstruction and usually present in neonatal age soon after birth [1]. However, delayed presentation has been documented in literature in first few months or years of life related to partial obstruction. [2]. Here we report duodenal web presenting in the third year of life associated with trichobezoar in the duodenum.

A 2-year-old girl with Down syndrome presented with complaints of recurrent vomiting of 2-3 months. The vomiting was non-bilious in nature and it contained 'cherry seeds' eaten about 3-4 months before. The child had no history of abdominal distension, blood in vomitus or bowel complaints. The child was well hydrated, afebrile, and with no previous complaints. Abdominal examination showed non-distended abdomen. No definite lump or tenderness was palpable. There was no free fluid and bowel sounds were normal. *X*-ray of abdomen revealed 'double bubble' with paucity of distal gas. A contrast study was done using water-soluble contrast agent, which showed hugely distended stomach with delayed drainage and normal small bowel. She was explored through supra-umbilical transverse incision. A hugely dilated stomach was identified. The second part of duodenum had a windsock deformity. Duodenotomy revealed a pre-ampullary web with trichobezoar obstructing the lumen. The gastric outlet was normal. The child was managed by duodenoduodenostomy. She remained well in post-operative period and is well on follow-up after 3 months.

Duodenal atresia or duodenal web may be identified on antenatal ultrasound or usually presents in first week of life with recurrent vomiting. The condition can be picked up on plain x-ray of abdomen showing 'double-bubble' appearance. Duodenal web; however, may present late due to partial obstruction [1]. Duodenal atresia and duodenal web are caused by abnormal duodenal development at 6-8 weeks of gestation, and are known to be associated with Down syndrome [1].

Trichobezoar is a condition where a collection of hairs form a mass that does not pass into the intestine and causes obstruction. Usually the trichobezoar occurs in the stomach, and it may extend into the intestine as a tail causing Rapunzel syndrome [3]. The trichobezoar occurs more commonly in persons with psychiatric diseases with trichotillomania. Although no behavior of eating hair was noted by parents, the same may be present/ have happened accidentally due to intellectual deficit or inadequate supervision by parents. The cherry seeds reported in history may have precipitated the obstruction either by themselves or by acting as a nidus for the trichobezoar. Trichobezoar is usually diagnosed on ultrasound or CT scan of abdomen, and managed by retrieval through laparotomy or laparoscopy [4].

A similar case has been reported in the French literature [5]. The association of duodenal atresia and Down syndrome helped us in suspecting the duodenal atresia. Although phytobezoars have been reported in early life, the incidence of trichobezoar in third year of life is rare. For a child with Down syndrome and recurrent vomiting, the differential for duodenal atresia should be high on the list and needs to be evaluated and managed promptly.

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CASPR2-Mediated Autoimmune Encephalitis in a Toddler

Autoimmune limbic encephalitis commonly presents in adults, frequently associated with Leucine-rich glioma inactivated protein-1 (LGI1) and Contactin associated protein-2 (CASPR2) antibodies, and is uncommon in children. Here we report a case of CASPR2 mediated autoimmune limbic encephalitis in a toddler.

A 19-month-old girl, born to consanguineously married parents, who was developing typically, presented with excessive irritabilityand decreased sleep for the past 40-50 days.Child had received diphtheria-pertussis-tetanus(DPT) booster dose 50 days back. Initially the symptoms were attributed to vaccination, but irritability kept gradually increasing. Child gradually regressed in language and cognition. She was not able to speak and on admission was able to only coo.There was behavioural change in the child in the form of new onset head banging, throwing of previously favourite toys and was avoiding going to the mother.Child was not mingling well

with others, did not play like before, was not showing interest in surroundings and there was cognitive decline in the form of not showing body parts and not recognizing mother. However, child was able to walk without support, and there was no motor regression. There was no history of seizures, fever, vision disturbance, involuntary movements, abnormal eye movements, and ataxia. Child had an uneventful birth history and family history. On examination, child was extremely irritable and crying inconsolably. Child had pallor and vitals were stable. There was no lymphadenopathy and organomegaly. There were dried crusted rashes with excoriation over trunk and limbs, with hyperpigmentation. Neurological examination did not show any cranial nerve involvement or motor weakness. There was no muscle twitching/fasciculation/neuromyotonia with spasticity, the best elicited power was 4/5 in all limbs,with brisk deep tendon reflexes with ankle clonus with bilateral extensor plantar response. Sensory and cerebellar examination was normal. Meningeal signs were absent.

Possibility of autoimmune encephalitis, connective tissue disorder and malignancy, possible lymphoma with paraneoplastic manifestations were considered as differential diagnosis. Peripheral smear examination, erythrocyte sedimentation rate and C-reactive protein were within normal units, and serum antinuclear antibody was negative. Magnetic resonance imaging of brain and nerve conduction study were normal. CSF analysis revealed one lymphocyte, normal protein and sugar, and positive oligoclonal bands. Electroencephalogram revealed diffuse slowing. Autoimmune encephalitis panel done by indirect immunofluorescence technique in transfected cells was positive for CASPR2 antibody in the serum and negative in the CSF.

Chest *X*-ray, ultrasound abdomen, computed tomography (CT) chest, CT abdomen, and bone marrow aspiration done to rule out the possibility of malignancy were non-contributory. Child was treated with intravenous methylprednisolone for 5 days with intravenous immunoglobulin 2g/kg, and showed clinical improvement within 72 hours. The child's irritability decreased and gradually child started sleeping well, started showing interest in surroundings, regained previous vocabulary of nearly 100 words, speaking two word phrases and was interacting well. The skin lesions resolved within 10 days after starting immunosuppressive therapy. After administration of intravenous methylprednisolone for five days, child was treated with oral prednisolone 2 mg/kg bodyweight per day for 6 months and was gradually tapered and stopped as child was completely asymptomatic and was gaining new milestones. Child has been on follow-up for the last one year. At age of 29 months, the child has achieved age-appropriate developmental milestones.

CASPR2–mediated limbic encephalitis is characterized by the onset of cognitive deficits, psychiatric disturbances, seizures, peripheral nerve hyper-excitability, neuropathic pain and insomnia in association with detection of Caspr2 antibodies in serum or cerebrospinal fluid, with or without underlying malignancies. There is strong male predominance with risk of malignancy in adults [3].

CASPR2 is a membrane protein expressed in the central and peripheral nervous system, which is essential for proper localization of voltage-gated potassium channels (VGKC) [4]. The most common presenting symptoms in adults are cognitive disturbance (26%), seizures (24%), peripheral nerve hyperexcitability (21%) and neuropathic pain (18%) [3]. Fewer than 10 pediatric cases have been reported with CASPR2 autoimmunity, so the phenotypes and immunotherapy responsiveness is less well-defined in children [4]; though, encephalopathy, seizures, neuropsychiatric symptoms, neuropathic pain and cramps are described as clinical features in children, with a median age of onset of 13 years [5,6]

Children with CASPR2 autoimmunity are under-recognized because neuropathic pain and symptoms of peripheral nerve hyperexcitability are difficult to characterize in children [5]. Peripheral nerve hyperexcitability has been documented in patients with CASPR2 autoimmunity[1]. One of the limitations of our case report is that we could not document the presence of neuromyotonia or evidence of peripheral nerve hyperexcitability in this child by electrophysiology.

The sub acute onset of behavioral disturbance, cognitive and speech regression should be considered as clinical clues to suspect autoimmune encephalitis in children; and CASPR2 mediated autoimmune limbic encephalitis should be considered as a cause of irritability with intractable itching/neuropathic pain in a child, as its recognition is important because they respond well to immunosuppressive therapy.

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