Acinar cell tumour of pancreas in a 4 year old child

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Abstract

Abstract: Paediatric pancreatic tumours by virtue of their rarity pose a significant challenge for the management.1 Although pancreatoblastoma and acinar cell carcinoma are distinct tumour entities, there is considerable overlap of histopathological characteristics which makes the diagnosis difficult pathologically.2 In infancy and childhood pancreatoblastoma is the most common tumour.3 Acinar cell carcinoma usually presents in older children.4,5 We describe a four year old child with acinar cell carcinoma which is possibly only the fourth case with this condition at such a young age.

Key words:

Acinar cell carcinoma; pancreatic tumour; pancreatoblastoma.

Introduction:

Pancreatic tumours are rare and pose a significant challenge to the paediatric surgeons and pathologist.[2] In order to ensure good outcome complete surgical resection is necessary.[6,7] The child might also have to be subjected to neoadjuvant chemotherapy in order to ensure complete surgical removal.[8] The chemo sensitivity and radio sensitivity is a debatable issue but lack of any standardized protocol makes it necessary to individualize the management.[9]We describe here a child diagnosed as pancreatoblastoma, who had unresectable disease but responded well to chemotherapy and complete resection was possible. On postoperative histopathology it turned out to be acinar cell carcinoma which is a rare tumour especially in this age group.

Case summary :

A four year old child presented with history of fall during playing one day back followed by pain in abdomen epigastric region. He had non bilious vomiting. Mother gave history of progressive distention of abdomen since last two months. Child also had transient jaundice 2 months back. There was no history of fever. On examination there was a large lump palpable in epigastric and umbilical region.

Ultrasound revealed large heterogeneous mass in head of pancreas with suspicious breach of capsule and hemoperitoneum. There was infiltration into the root of mesentry. Tumour caused widening of 'C' loop of duodenum with displacement of 2nd and 3rd part of duodenum. Diagnosis of pancreatic tumour possibly pancreatoblastoma was considered. CT scan confirmed the findings. The tumour was large heterogenous and arising from head, neck and uncinate process of pancreas with size of 8.4 x 9.7 x 9.9 cm. The mass was reaching up to anterior abdominal wall and posteriorly compressing Inferior vena cava. It was encasing portal vein at its origin. There was breach in capsule. CT chest showed mild bilateral pleural effusion. Serum alpha fetoprotein was raised (12170.87 ng/ml).CT guided biopsy revealed tumour arranged in sheets and nests, expressing Creatine Kinase. The cytopathological features were consistent with pancreatoblastoma.

After initial management of pain and stabilization patient received chemotherapy, Cisplatin and Doxorubicin (PLADO). After 4 cycles of PLADO the tumour size decreased to 6.5 x 6.4 x 2.6 cm and previously seen encased portal vein was now free from tumour. The child was taken up for surgery. The tumour was situated

in head of pancreas. (Fig 1). The Superior mesenteric vein and portal vein were free. (Fig 2). The tumour could be excised with a margin of compressed pancreatic tissue around it. (Fig 3) There was a small breach in capsule during removal with resultant spill of tumour locally. Postoperatively patient recovered well. Patient was tolerating full orally by 5thpostoperative day. On 14th postoperative day patient developed pain in abdomen. Serum Lipase was raised (2190.5 U/lit) and ultrasound revealed heterogeneously echogenic cystic lesion in head of pancreas 3.7 x 4.1 cm size with perilesional edema. The child responded well to conservative management and serum Lipase decreased along with cystic lesion. Postoperatively the serum Alpha-fetoprotein decreased to 11 ng/ml. The child is now one month postop and is scheduled to receive abdominal radiation.

The histopathological report showed malignant epithelial tumour showing acinar differentiation. Margins were tumour free with compressed benign pancreatic tissue seen around the tumour. The pathology report had a comment that the earlier diagnosis of pancreatoblastoma was made considering the tumour cell characteristics and the age of the child. However absence of squamous rest in the multiple sections studied rule out the possibility of pancreatoblastoma. Possibility of neuroendocrine cell tumour was also ruled out by absence of synaptophysin and CD 56 expression and insignificant chromogranin expression.

Discussion:

The pancreatic tumours in paediatric population are rare. The incidence of malignant pancreatic tumours in children was 0.18 per 1,00,000 people in United State of America in 2008.[5] The tumours may be endocrine, exocrine or sarcomas.[10,11] The rarity of these tumours limit our understanding of the pathophysiology and the clinical course and hence the appropriate management of the tumours. There are no standardised protocols and hence the management has to be individualised according to the given conditions.[9]

Among the paediatric pancreatic malignancies pancreatoblastoma is the most common tumour in children less than 10 year of age. The usual age of presentation is 4- 5 years.[3] They usually tend to present as large abdominal masses followed by pain in abdomen.[12] Serum alpha feto protein is elevated in 68% to 94% of patients. Although it is of no diagnostic value, it has utility in terms of marker of response to chemotherapy and of recurrence in postoperative period.[8] The treatment is complete resection of the lesion.[13] The resection may include excision, pancreaticoduodenectomy, and distal pancreatectomy depending on the site of the lesion. For unresectable lesions neoadjuvant chemotherapy in the form of PLADO is generally recommended as these tumours have shown sensitivity to this regimen.[8,9]Histopathologically they exhibit dense cellularity with characteristic "squamoid corpuscles".[2] The 15 year survival rate in children is reported to be 61%.[5]

Acinar cell carcinomas are rare pancreatic neoplasms accounting for 1-2% of all pancreatic malignancies. In children the incidence is 7.2% of all pancreatic malignancies.[14] Acinar cell carcinomas are generally seen in children more than 10 years of age.[15] The most common site for acinar cell carcinoma is tail (41% of cases) followed by head (32% of cases) of the pancreas. They usually present with large palpable masses and abdominal discomfort.[14] Grossly they have circumscribed expansile growth pattern and invasion of common bile duct is less frequent. Jaundice is rarely seen.[12] The classic "Lipase hypersecretion syndrome" has not been observed in children.[16] Serum alpha feto protein has been observed to be elevated in all patients of acinar cell carcinomas. It has been hypothesized that alpha feto protein production in pancreatic tumours is related to acinar differentiation.[6] Acinar cell carcinoma in children has overlapping features with pancreatoblastoma and may closely resemble neuroendocrine tumours. Squamous nests which are absent in acinar cell carcinoma are considered the feature essential for the diagnosis of pancreatoblastoma.[2]

Although pancreatoblastoma is predominantly a tumour of infancy and childhood and acinar cell carcinoma is of adults, exceptions to the above are well documented.[17,18] Acinar cell carcinoma occurring in childhood has considerable clinical as well as pathological overlap making the diagnosis difficult. Often the diagnosis is made by pathologist on the basis of the age of the patient.[18]In our patient the same has happened. A thorough literature search revealed only three cases of acinar cell carcinoma in 4 year old child and less.[19, 20] To the best of our knowledge this is only the fourth case of acinar cell carcinoma in such a young child. The preoperative diagnosis in our patient was pancreatoblastoma and as imaging features suggested the non

resectability the child was subjected to neoadjuvant chemotherapy to which he had responded. Surgically the resection was satisfactorily complete with negative margins. Radiotherapy has limited role but as our child had tumour spill he is scheduled for the radiotherapy. As mentioned earlier the therapy has to be individualized for the best outcome.

Conclusion:

Paediatric pancreatic tumours are rare. Acinar cell is a rare type of pancreatic tumour with vast differences between its adult counterpart in terms of clinical presentation, management and histopathological features. Rarity of this tumour type results in inability to frame a standard management protocol. Complete surgical resection is associated with good prognosis.

Conflict of interest: None

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Fig 1: Tumour in head of pancreas with compressed pancreatic tissue around it

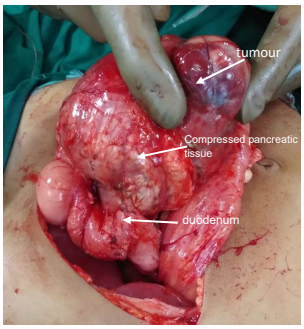

Fig 2: Tumour free from superior mesenteric vein

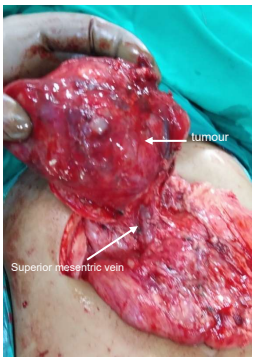

Fig 3: Excised specimen