Letter

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# Pancreatic lipomatosis in cystic fibrosis: Rare manifestation of an uncommon disease

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**Summary** **Cystic fibrosis is deemed to be uncommon in India. The presentation is usually in the childhood although more cases are now being recognized in adolescence and adulthood. We report a case of an adolescent male who had been treated for recurrent pulmonary infections and received anti-tubercular therapy for a possible diagnosis of sputum negative pulmonary tuberculosis and was evaluated for steatorrhea. The presence of pancreatic exocrine insufficiency along with pancreatic lipomatosis suggested the diagnosis of cystic fibrosis.**

***Keywords:*** Cystic fibrosis, bronchiectasis, pancreatic exocrine insufficiency, steatorrhea, endoscopic ultrasound

Cystic fibrosis is a multisystem disorder characterized by recurrent pulmonary infections, pancreatic exocrine insufficiency and is usually diagnosed in young age. It is an inherited disorder and the age and manner of presentation vary depending on the severity of defect in the cystic fibrosis gene. Since a large number of mutations have been reported the clinical presentation is extremely variable (*1*). We report about an 18 year old male who had been treated as sputum negative tuberculosis elsewhere and presented to us with steatorrhea prompting evaluation for cystic fibrosis.

An 18-year old male had been symptomatic for recurrent episodes of cough associated with mucoid expectoration and occasional hemoptysis for 7 years. He had been treated with multiple courses of antibiotics and had received 6-month therapy for a suspected diagnosis of sputum negative pulmonary tuberculosis elsewhere. For the past 6 months, he had noticed an increased bulk of his stools and passage of oily stools. This was associated with significant loss of weight (6 kg) and had developed temporal hollowing suggesting fat malabsorption. He denied any history of abdominal pain. To avoid steatorrhea, he had reduced the intake of fat in his meals. His body mass index was 15.6 kg/m$^2$. The patient's younger sibling had died of an undiagnosed respiratory illness at 4 years of age. His parents were asymptomatic and did not report any respiratory illness. His fecal elastase levels were done for the possibility of pancreatic exocrine insufficiency and were 53 (normal > 200 μg/g of stool). His abdominal and chest computed tomography revealed a hypo-attenuating pancreas (Figure 1A) consistent with complete pancreatic lipomatosis and evidence of bronchiectasis (Figure 1B). Endoscopic ultrasound confirmed the presence of diffusely hyperechoic pancreas consistent with pancreatic lipomatosis (Figure 1C). The sweat chloride levels were elevated at 79 mmol/L. This was repeated

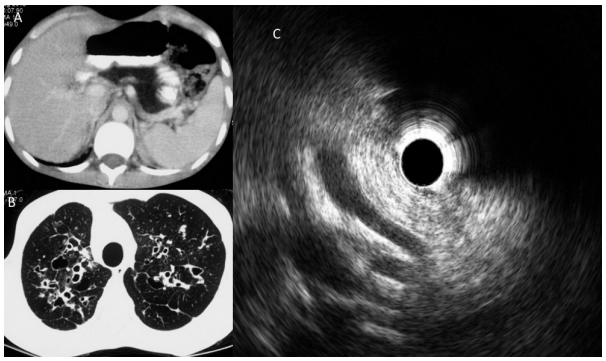


**Figure 1. (A), CT abdomen showing pancreatic lipomatosis; (B), CT chest showing bronchiectasis; (C), Endoscopic ultrasound showing hyperechoic pancreas.**

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**Table 1. Showing results of diagnostic investigations**

| Feature | Investigation | Findings |
|---|---|---|
| Pancreatic involvement | Computed tomography | Pancreatic hypoattenuation |
| | Endoscopic ultrasound | Hyperechoic pancreas |
| | Fecal elastase | Low levels |
| Pulmonary Involvement | Chest X Ray | Bronchiectasis |
| | Computed tomography | Bronchiectasis |
| Basis of diagnosis | Bronchiectasis | |
| | Pancreatic exocrine insufficiency | |
| | Elevated sweat chloride | |
| | Pulmonary disease in sibling | |

after one month and the levels were still elevated. Therefore, a diagnosis of cystic fibrosis with pulmonary and pancreatic involvement with pancreatic lipomatosis was made. The patient was started on pancreatic enzyme supplementation along with twice daily proton pump inhibitor. The patient reported improvement in steatorrhea and at 3 months of follow-up reported a gain of 4 kg of his weight. He has been advised to continue follow-up for his pancreatic insufficiency

Cystic fibrosis, although reported as case reports, is believed to be uncommon in India (*2*). Given the high prevalence of pulmonary tuberculosis, it is not unexpected that cystic fibrosis may be confused with pulmonary tuberculosis especially with an adolescent presentation and absence of other organ system involvement as in our case. The appearance of steatorrhea directed the evaluation to pancreatic pathology and brought to light the diagnosis in a patient who had evidence of underlying bronchiectasis (Table 1).

Interestingly, the patient also had complete pancreatic lipomatosis which by itself is uncommon and is variously described as pancreatic lipomatosis or steatosis or adipose atrophy. The fatty replacement may involve a part of the pancreas or may be total, as in the present case (*3*). While in adults the causes may include diabetes mellitus, insulin resistance, obesity, chronic pancreatitis, pancreatic ductal obstruction due to stone or malignancy, in younger age group certain hereditary conditions like cystic fibrosis and Shwachman-Diamond syndrome may be responsible (*4*). Although the entity is uncommon, in patients with cystic fibrosis the presence of complete pancreatic lipomatosis and partial lipomatosis has been reported to be as high as 41% and 19% in one report (*5*). To conclude, the presence of pulmonary symptoms and bronchiectasis along with pancreatic exocrine insufficiency must prompt evaluation for cystic fibrosis.

## References

1. Fajac I, De Boeck K. New horizons for cystic fibrosis treatment. Pharmacol Ther. 2017; 170:205-211.
2. Ashavaid TF, Raghavan R, Dhairyawan P, Bhawalkar S. Cystic fibrosis in India: A systematic review. J Assoc Physicians India. 2012; 60:39-41.
3. Ambesh P, Lal H. Pancreatic lipomatosis: Complete replacement of pancreas by fat. J Clin Diagn Res. 2015; 9:OL01.
4. Anand R, Narula MK, Chaudhary V, Agrawal R. Total pancreatic lipomatosis with malabsorption syndrome. Indian J Endocrinol Metab. 2011; 15:51-53.
5. Feigelson J, Pécau Y, Poquet M, Terdjman P, Carrère J, Chazalette JP, Ferec C. Imaging changes in the pancreas in cystic fibrosis: A retrospective evaluation of 55 cases seen over a period of 9 years. J Pediatr Gastroenterol Nutr. 2000; 30:145-151.

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