

lepto-meningeal melanocytosis resulting occasionally in the development of hydrocephalous, seizures and leptomenigeal melanoma(5,7).

Other associated abnormalities include spina bifida or meningocele (when the lesion is over the spine), club foot, hypertrophy or atrophy of the deeper structures of a limb(5,7). These associated anomalies were not observed in the present case, though the skin over the spine was involved. The course is fairly stable in most cases as has been observed in the present case. They may elevate with time, change texture, develop irregular nodularity, variably darken or lighten. Lesions very rarely regress.

Giant congenital melanocytic nevus runs a significant risk of malignant melanoma in 6-10% of the cases(7). Because of the high risk of malignant degeneration during childhood early excision has been recommended(2,4). Aggressive surgical excision early in life is often not practical because of involvement of vital structures as well as the functional and cosmetic disability caused by excising large cutaneous areas.

It is recommended to follow up patients every 6-12 months interval to observe for changes like asymmetry, border irregularity, localized color variation to red, dark brown, blue or black, changing size of atypical area and for appearance of elevated firm nodules(6).

REFERENCES

1. Jacobos AH, Walton RG. Incidence of birth marks in neonate. *Pediatrics* 1976, 58: 218-222.
2. Monique RE, Grant-kels MJ. Important melanocytic lesions in childhood and adolescence. *Pediatr Clin North Am* 1991, 38: 791-809.

3. Rhode AR, Melski JW. Small congenital nevovellular nevi and the risk of cutaneous melanoma. *J Pediatr* 1982, 100: 219-222.
4. Kaplan EN. The risk of malignancy in large congenital nevi. *Plast Reconstr Surg* 1974, 53: 421-428.
5. Mackie MR, Melanocytic nevi: Textbook of Dermatology, 3rd edn. Eds Rook A, Wilkinson DS, Ebling FJG. Oxford, Blackwell Publications, 1979, pp 179-183.
6. Eichenfield LF, Honigs PJ. Difficult diagnostic and management: Issues in pediatric dermatology. *Pediatr Clin North Am* 1991, 38: 687-710.
7. Esterly BN. The skin-cutaneous nevi. In: Nelson Textbook of Pediatrics, 13th edn. Eds Behrman RE, Vaughan VC. Philadelphia, WB Saunders Co, 1987, pp 1393-1395.

Esophagopleural Fistula Complicating Suppurative Lung Disease

V. Kohli

R.K. Marwaha

K.L. Narasimhan

R.P.S. Bajwa

S.K. Mitra

Acquired esophagorespiratory fistulas are rare in childhood. They may be esophagobronchial or esophagopleural, the

From the Departments of Pediatrics and Pediatric Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

Reprint requests: Dr. R.K. Marwaha, Additional Professor, Department of Pediatrics, PGIMER, Chandigarh 160 012.

former being more common(1). The symptoms are often non-specific and the diagnosis delayed. Considerable controversy regarding optimal treatment still exists(2,3) and recommendations range from conservative supportive management to primary closure. The rarity of reports of esophagopleural fistulas in children has prompted us to describe such a case in a young girl.

Case Report

A 6-year-old girl, presented with 20-day history of cough, fever and chest pain. Clinical features were consistent with a right sided pleural effusion. An intercostal tube with underwater seal drain was introduced and 150 ml thick pus drained. *Staphylococcus aureus* was isolated from the pus and appropriate antibiotics started. In spite of fifteen-day treatment with parenteral cloxacillin and gentamicin and intercostal drainage, there was no significant improvement. The patient was subjected to a thoracotomy and debridement of the chest. About 200 ml of pus was present in the thoracic cavity. A thin pleural peel over the visceral pleura was removed to allow lung expansion. On the second post-operative day ingested food material was seen to be draining from the chest tube. A similar observation, a day prior to surgery, of rice grains being present in the drainage tube had been disregarded. The presence of an esophagopleural fistula was confirmed by a barium swallow, the origin being in the lower third of the esophagus. Endoscopy revealed the rest of esophagus to be healthy. The patient was then started on isonex, rifampicin and pyrazinamide. There was, however, no evidence of tuberculosis on histopathological examination of pleural biopsy. The culture of pleural pus of mycobacteria was also negative. In view of the

poor general condition of the child, she was managed conservatively and underwent a feeding jejunostomy as nasogastric feeding aggravated drainage from the chest tube. The subsequent course was complicated by colonization of the pleural cavity by *Candida albicans*. Jejunostomy feeding ensured a weight gain of 4 kg over the next 3 months. She had a persistent bronchopleural fistula in the post-operative period and was discharged on open drainage.

Repeat radiological studies, 3 months later, showed a persistent fistula (Fig. 1A). Thoracotomy done, at this instance, revealed a fistula of 5 mm size at the lower third of the esophagus. The fistula was closed with 50 vicryl interrupted stitches and densely adherent to the chest wall and diaphragm. The rest of the esophagus was normal. The post-operative recovery was uneventful and oral feeds were started after a contrast swallow study showed no leak (Fig. 1B). The feeding enterostomy tube was removed on the tenth post-operative day. Antitubercular treatment was continued for a total period of 9 months. She is now able to comfortably carry out routine physical activity even though her right lung is only partially expanded. The open chest tube drainage was removed after drainage decreased to negligible amounts.

Discussion

The patient described had an esophageal perforation with empyema and an associated lung disease. Esophageal rupture or perforation is usually an iatrogenic, life-threatening complication secondary to endoscopy or esophageal dilatations(2,4). Esophagorespiratory fistulas, on the other hand, follow esophageal or paraesophageal infections(1). Caseating mediastinal lymph nodes may simultaneously erode into the

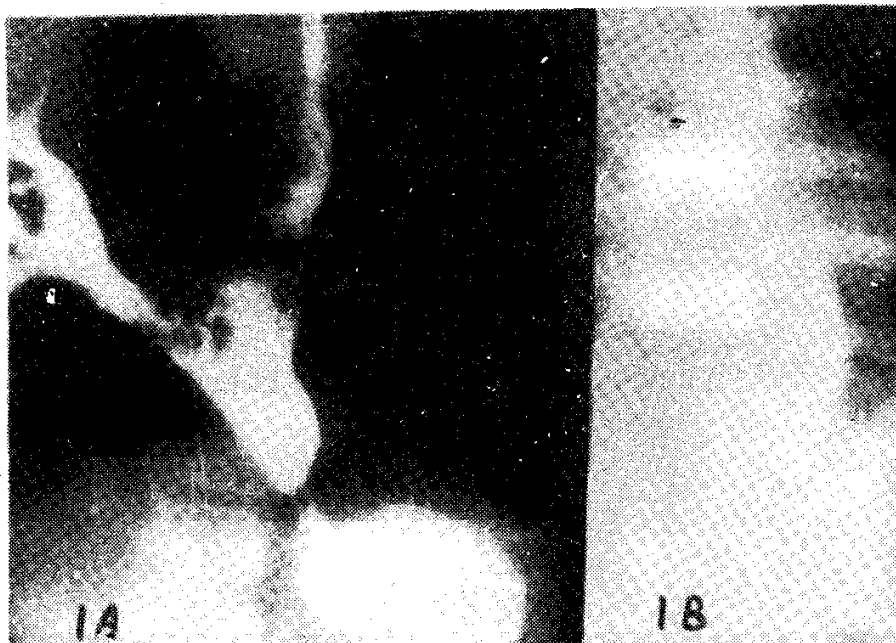


Fig. 1. Barium swallow showing the fistulous leak in the lower third of the esophagus, showing (A) persistent communication after 20 weeks, (B) post-operative contrast study showing no leak.

esophagus and airway (trachea or main bronchus) or the pleural cavity leading to a fistulous communication. Tuberculosis is the commonest etiological disease although esophageal candidiasis and other fungal diseases such as actinomyces and blastomycosis are also reported(1). The etiology for the fistula in our patient is not clear, although the therapeutic response to antitubercular drugs suggests a tuberculous etiology.

The diagnosis of an esophagorespiratory fistula may be delayed for weeks to several years(1). The triad of tachypnea, abdominal rigidity and subcutaneous emphysema described initially by Barret(5) in cases with spontaneous rupture of the esophagus has been shown to be uncommon in subsequent reports(2,4). In our patient, the presence of an underlying esophageal perforation remained hidden till the empyema cavity was debrided and food particles appeared in the drainage

bottle. A fistula should be suspected if the patient coughs on swallowing liquids. The diagnosis can be established at the bed side by the methylene blue test. Esophagoscopy localises the site of the fistula and detects a primary esophageal infection if present. Contrast studies with gastrografin define the tract and are a prerequisite before surgery.

The principles of treatment in esophagorespiratory fistula are similar to those presently adopted for esophageal rupture or perforation. Management is non-operative in cases where the diagnosis has not been established within 24 hours(2-4). Conservative management includes nasogastric suction, adequate pleural drainage, broad-spectrum antibiotics and either a feeding enterostomy or total parental nutrition. Cimetidine has been advocated to promote healing by reducing acid reflux(6). Esophageal 'rest' achieved in this manner, for periods of weeks to

months coupled with specific therapy directed at the etiology, may result in spontaneous closure of the fistulous communication. A persistent leak, as in our case, may necessitate a delayed surgical repair. An alternate, relatively more aggressive approach has been recommended by some authors and involves performing a resection and immediate restoration of continuity of the gastrointestinal tract, but is associated with a higher mortality(2).

REFERENCES

1. Kramer P, Burakoff R. Infections of esophagus. In: Bockus Gastroenterology, Vol 2. Eds Haubrich WS, Kalser MH, Roth JLA, Schaffner F. Philadelphia, WB Saunders Company, 1985, pp 787-800.
2. Bladergroen MR, Lowe JE, Postlethwait RW. Diagnosis and recommended management of esophageal perforation and rupture. *Ann Thorac Surg* 1986, 42: 235-239.
3. Walker WS, Cameron EWS, Walbaum PR. Diagnosis and management of spontaneous transmural rupture of the esophagus. *Br J Surg* 1985, 72: 204-207.
4. Lyons WS, Seremetis MG, de Guzman VC, Peabody JW. Ruptures and perforations of the esophagus: The case for conservative supportive management. *Ann Thorac Surg* 1978, 25: 346-350.
5. Barret NR. Spontaneous perforation of the esophagus: Review of literature and Report of three new cases. *Thorax* 1946, 1: 48-70.
6. Ivey TD, Simonowitz DA, Dillard DH, Miller DW. Boerhaave syndrome: Successful conservative management in three patients with late presentation. *Am J Surg* 1981, 141: 531-533.