Novel treatment (new drug/intervention; established drug/procedure in new situation)

A staged management of prolonged chylothorax in a patient with yellow nail syndrome

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Summary

Spontaneous chylothorax remains an unusual condition with diverse aetiologies and non-unified management strategies. Owing to the rarity of the condition, the evidence of management remains from case reports and limited retrospective studies. This case represents a difficult-to-manage chylothorax secondary to yellow nail syndrome with initial failure of both surgical and conservative treatment methods. Pleurovenous shunting represents a surgical management approach allowing the patient to live with and control an ongoing chyle leak.

Background Yellow nail syndrome is a rare disease formed by the clinical triad of pleural effusion, lymph oedema and yellow nails.^{1–4} Massive chylous pleural effusion is often the most challenging aspect of the condition.⁵

CASE PRESENTATION

A 72-year-old man was admitted to a teaching hospital in January 2011 and treated for a right-sided community-acquired pneumonia with parapneumonic effusion (pleural fluid analysis—pH 7.49, protein 21 g/l, pus cells ++, no bacterial growth and inflammatory cells only seen). Three litres of effusion were drained. He was discharged to be followed up in the clinic in February, however, was readmitted later that month with shortness of breath. On examination he was not feverish with a respiratory rate of 28 and stony dullness across the right side of the chest. Chest radiograph showed a massive right-sided pleural effusion with meniscus sign (figure 1). On insertion of an intercostal drain 7 litres of chylous fluid were drained (pleural fluid analysis—triglyceride 2.46 mmol/l and cholesterol 0.4 mmol/l). A CT scan showed no evidence of malignancy or no anatomical explanation for the chyle leak. After drainage the patient was again discharged.

Having been discharged the patient was readmitted again in late February with a recurrence of the right-sided chylothorax (fluid—triglyceride 2.82 mmol/l and cholesterol 0.6 mmol/l). At this point he was started on a low-fat diet and transferred to the thoracic surgical team on suspicion of a leak from the thoracic duct.

Given the nature of an ongoing unresolved chylothorax after a failure of conservative management, the patient was scheduled for a right thoracotomy to visualise the thoracic duct with a view of potentially oversewing any identified leak or ligation of the duct if necessary. Intraoperatively, a leak from the duct was clearly visualised (cream was given preoperatively to aid visualisation of the duct), the duct was tied off six times and clipped, a surgical pleurectomy was performed and two intercostal drains were left in situ. During the initial 5 days of the postoperative period, the total drain output remained at

between 500 and 950 ml of yellow serous fluid while the patient was on a strictly fat-free diet. In view of persistent high total drain output, octreotide was started but the drain output continued at 200–800 ml daily. On post-operative day 16, a trial of normal diet was started, as the fluid had remained clear and non-chylous. Once on a normal diet, 900 ml of chylous milky fluid was drained the following day, and therefore clinically the patient had an ongoing chylous leak.

Previous to this admission, a diagnosis of yellow nail syndrome had not been made. On closer enquiry, yellowing of the nails was reviewed on examination. On this basis of the clinical triad of yellow nails, chronic lymphoedema and chylous pleural effusion, a diagnosis of yellow nail syndrome was made. Microscopy and culture of samples from the yellowed nails excluded fungal infection.

In view of the failed surgical ligation and failure of conservative management, lymphoscintigraphy was performed to assess the lymphatic anatomy in view of possible percutaneous embolisation or pleuroperitoneal shunt.

Figure 1 Chest x-ray on presentation.

The lymphatic system was demonstrated to be severely abnormal, especially on the left side. No tracer uptake was observed within the thoracic cavity. Hence, percutaneous embolisation was not possible and the lymphatic system was judged as not being able to support significant amounts of fluid being shunted into the abdominal cavity.

In view of the ongoing large effusion, the decision was taken jointly with the patient to return to the theatre for a pleurovenous shunt. A Denver-type shunt (figure 2), consisting of a catheter sitting in the pleural space, connected to a one-way valve with 1.5 ml pump chamber (palpable subcutaneously), allowing the fluid to be manually pumped through and into the right subclavian vein. The shunt was inserted via three incisions and great care was taken to ensure no air was introduced into systemic circulation (figure 3). The shunt functioned well and there were no immediate postprocedure complications.

INVESTIGATIONS

- ▶ Pleural fluid analysis
- ▶ Chest radiograph
- ▶ CT scan
- ▶ Nail microscopy and culture
- ▶ Lymphoscintigraphy

DIFFERENTIAL DIAGNOSIS

- ▶ Cholesterol pleural effusion
- ▶ Malignant pleural effusion
- ▶ Infective pleural effusion

TREATMENT

- ▶ Intercostal drain
- ▶ Surgical ligation and pleurectomy
- ▶ Fat-free diet
- ▶ Octreotide
- ▶ Pleurovenous shunt

OUTCOME AND FOLLOW-UP

The patient made good recovery postoperatively, the shunt has remained to function well and has controlled the effusion. The patient has been required to manually pump the shunt 300–400 times daily, this has proven effective and well tolerated.

Figure 2 Denver shunt before insertion.

Figure 3 Intraoperatively during shunt insertion.

In the 4 months following shunt insertion the patient has been readmitted twice to the hospital for unrelated problems (sepsis and a flare of encephalitis), on both occasions intercostal drains have been required as the patient has been too unwell to operate the shunt. Following discharge the shunt has remained fully effective (figure 4).

DISCUSSION

Prolonged chylothorax is a difficult scenario for both patients and clinicians. Owing to the rarity of the problem, the up-to-date evidence of management is derived from retrospective rather than prospective data.⁶ Radiation

Figure 4 Chest x-ray postprocedure at follow-up.

therapy appears to have some success in patients having chylothorax who have an underlying malignancy.⁷ Failing that, pleurodesis using Talc is a further option.⁸ Yellow nail syndrome itself is also a rare diagnosis. We have only been able to find one similar case in the literature.⁹ Where the use of a pleuroperitoneal shunt in a patient of yellow nail syndrome has been tried.⁵ Surgical treatment, such as pleurectomy and oversewing of leaking collaterals, are reserved for symptomatic and persistent cases in general. Stringently selected patients may benefit from the implantation of pleuroperitoneal or pleurovenous shunts. In terms of nutritional support, some clinicians consider a fat-free diet or medium-chain triglyceride diet. Recently, percutaneous embolisation of the thoracic duct is one of the newer modalities of intervention.¹⁰ In a systematic review, a group of 35 children suffering from chylothorax experienced some improvement after receiving octreotide or somatostatin.¹¹ Ongoing research to determine 'what is the best treatment' will aid our understanding of chylothorax. Owing to the lack of evidence, variations in management are unavoidable. Nevertheless, a staged management that progresses from conservative management to radiological or surgical intervention would be considered as a logical approach.

Learning points

- ▶ Massive ongoing chylothorax in patients with yellow nail syndrome is rare but can be potentially challenging to manage.
- ▶ Pleurovenous shunts can provide a satisfactory management of ongoing pleural effusions; however, there is a lack of published data of long-term outcomes.
- ▶ There is no prospective study to guide the management of prolonged chylothorax, therefore it is an area based on clinical judgement, but a stepwise approach is recommended.
- ▶ Extended-term multicentre retrospective study as well as case reports would continue to bridge the gap in knowledge.

Competing interest None.

Patient consent Obtained.

REFERENCES

1. **Doerr CH**, Allen MS, Nichols FC III, *et al.* Etiology of chylothorax in 203 patients. *Mayo Clin Proc* 2005;**80**:867–70.
2. **Valentine VG**, Raffin TA. The management of chylothorax. *Chest* 1992;**102**:586.
3. **Samman PD**, White WF. The 'yellow nail' syndrome. *Br J Dermatol* 1994;**76**:153–7.
4. **Emerson PA**. Yellow nails, lymphoedema, and pleural effusions. *Thorax* 1966;**21**:247–53.
5. **Brofman JD**, Hall JB, Scott W, *et al.* Yellow nails, lymphedema and pleural effusion. Treatment of chronic pleural effusion with pleuroperitoneal shunting. *Chest* 1990;**97**:743–5.
6. **Merrigan BA**, Winter DC, O'Sullivan GC. Chylothorax. *Br J Surg* 1997;**84**:15.
7. **Roy PH**, Carr DT, Payne WS. The problem of chylothorax. *Mayo Clin Proc* 1967;**42**:457.
8. **Mares DC**, Mathur PN. Medical thoracoscopic talc pleurodesis for chylothorax due to lymphoma: a case series. *Chest* 1998;**14**:731.
9. **Tanaka E**, Matsumoto K, Shindo K, *et al.* Implantation of a pleurovenous shunt for massive chylothorax in a patient with yellow nail syndrome. *Thorax* 2005;**60**:254–5.
10. **Cope C**. Management of chylothorax via percutaneous embolization. *Curr Opin Pulm Med* 2004;**10**:311.
11. **Roehrer CC**, Jung A, Proquitte H, *et al.* Somatostatin or octreotide as treatment options for chylothorax in young children: a systematic review. *Intensive Care Med* 2006;**32**:650.

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