Leukaemia-in-ano
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ABSTRACT
Fistula-in-ano is a very common surgical condition, caused by anal cryptoglandular inflammation. Most cases are idiopathic. Other causes such as Crohn’s disease, trauma and malignancy are well known. Management of fistula-in-ano is largely surgical, especially if the patient is symptomatic. The goal of surgical therapy is sepsis drainage, delineate anatomy and eradicate the fistula while preserving faecal continence. Establishing the aetiology is also crucial as often a combination of specialist medical therapy is required, for example, in Crohn’s disease. We report an extremely unusual case of fistula-in-ano on an elderly man with chronic lymphocytic leukaemia (CLL). Histology from the fistula track demonstrated CLL infiltration. This case, not previously reported on PubMed search, illustrates a good example of joint specialist medical (a haematologist) and surgical effort in successfully treating this symptomatic fistula-in-ano.

Case report
A 79-year-old Caucasian man was admitted as an emergency with perianal pain. He had a background of chronic lymphocytic leukaemia for over 40 years. After five years, he received brief treatment with chlorambucil and prednisolone for constitutional symptoms. Seventeen years later after his initial diagnosis of CLL the patient had an ischiorectal abscess, which was incised and drained. His white cell count was consistently around 70x10⁹ cells/litre at that time. The wound healed and there were no further consequences until he presented with another abscess from the same area 15 years later: it was again incised and drained successfully and he was discharged from surgical follow-up. Between the two episodes of the ischiorectal abscesses, the patient was not on any treatment for his CLL.

Another eight years later, his white cell count was maintained with his normal levels between 50 and 60x10⁹ cells/litre. The patient underwent examination under anaesthesia and an abscess was found at five o’clock position, where there was evidence of previous incisions. Further incision and drainage took place but this time he was to be followed up with an MRI scan six weeks later.

T1 weighted MRI of his perineum showed complex high intersphincteric ischiorectal fistula draining out onto the surface at five o’clock position, with an internal opening just at left medial to 12 o’clock position. T2 weighted images showed a small abscess intersphincterically (Figure 1). The patient was electively brought back to theatre for another examination of anorectum under anaesthesia. The external opening was identified at 4cm from anal verge at five o’clock position and this opening was dilated. The fistula track was curetted to remove the granulation tissue and obtaining histology specimen. A seton was placed to improve drainage.
Clinical follow-up around two months later was arranged with both the surgeon and his haematologist. The seton was felt to be comfortable with minimal drainage. Histology of the fistula showed significant leukemic infiltration into the epithelial lining of the fistula (Figure 2). The opinion with the haematologist was that given his age and his stable CLL, no acute therapy was required.

Figure 2 shows a low and high-power haematoxylin and eosin view of the CLL deposits. These are irregular lymphocytic deposits, composed of a monotonous population of small and round dark cells. The deposits are surrounded by a fibrotic stroma and adipose tissue. The absence of a mixed population of lymphocytes suggests this is not simply a reactive process. Further immunohistochemical staining (not shown here) illustrated that the lymphocytes expressed were CD20 and CD23 and CD5 antigens positive, which is characteristic for CLL.

Discussion

Fistula-in-ano is considered a result of anal cryptoglandular inflammation. Most cases are idiopathic but occasionally can be consequent to a chronic inflammatory condition such as Crohn’s disease or tuberculosis. Seldomly, it results from invasion of local malignancy. Therefore, recurrent ischiorectal abscesses need to have a fistula, especially possible complex fistulae, either confirmed or excluded.
This gentleman had his first abscess drained in 1996. A possible recurrence was not until 2011. It would not have been unreasonable to consider the episode in 2011 to be a de novo abscess. Furthermore, the patient may not recall the event some 15 years earlier. CLL does have immunocompromising effect and the consequent development of an ischiorectal abscess would not be an unreasonable causative assumption similar to those with poorly controlled diabetes. This theory is nowadays challenged. The diagnosis of an underlying pathology should be established as it affects management of ischiorectal fistula.

The principle management of an ischiorectal fistula is to treat any sepsis, delineate the anatomy and eradication of the fistula. As with any chronic inflammatory conditions such as Crohn's disease or tuberculosis, eradication of the fistula often requires joint assistance of a medical specialist. In the case of Crohn's disease, once the sepsis is drained, gastroenterologists often commence on immunotherapy. Biopsies of the fistula track occasionally can help with establishing the diagnosis, assuring the medical specialists prior to commencement of these toxic medications.

Perianal complications resulting from haematological malignancy are seldomly reported. All the reported cases claimed the attribution to immunosuppression from either the primary disease or chemotherapy for the disease. Biopsies of our patient suggests leukaemic infiltration of the epithelial lining of the fistula. It is almost certainly unlikely that this recurrent or de novo fistula was directly caused by CLL. The more plausible theory is the immunosuppressive effects of CLL predispose to the activation or development of existing cryptoglandular inflammation. Furthermore, the presence of neoplastic tissue may have been all a while responsible for preventing healing of the fistula.

The opinion of the haematologist was to continue the watch and wait regime rather than commencement of immunotherapy such as Ibrutinib (tyrosine kinase inhibitor) and chemotherapy. In the case of mild CLL, watch and wait regime has demonstrated equivocal survival outcome for those considered asymptomatic. The debate from this case, which was controversial at the haematology multidisciplinary meeting, was that the patient had developed a fistula possibly consequent to or exacerbated by CLL. The consensus eventually was that given his age, free of symptoms and a maintained white cell count, risk of medical intervention may outweigh benefits. Radiotherapy of the anorectum was discussed. However, the patient is elderly with good fistula controlled of drainage, radiotherapy may worsen symptoms and, worse still, further prevent successful healing of the fistula.
Conclusion

Leukaemic infiltration within a fistula-in-ano has not been previously reported. The general principles of management of fistula in ano still applied. Importantly, this case illustrated that successful treatment of secondary complex ischiorectal fistula requires specialist multidisciplinary approach.

Competing interests:
Nil.

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