Sacrococcygeal Chordoma: A Rare And Easily Missed Diagnosis In Older Adults

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Abstract

Chordoma is a rare malignant primary bone tumour which can originate anywhere along the vertebral column. It has an incidence of 1 per million within the population in the United Kingdom with a preponderance for the male gender. It is caused by a mutation in a gene normally passed on in an autosomal dominant fashion within the healthy population. Most commonly these tumours affect the sacrococcygeal region (43-85%) and rarely can present like a pilonidal abscess. We present a case of a 68 year old female with an apparent pilonidal abscess which was found to be suspicious on incision and drainage. An invasive sacrococcygeal chordoma was diagnosed and resected. This case is one that highlights the importance of having a low index of clinical suspicion in older patients presenting with an apparent pilonidal abscess.

KEYWORDS: chordoma, pilonidal abscess
Abstract
Chordoma is a rare malignant primary bone tumour which can originate anywhere along the vertebral column. It has an incidence of 1 per million within the population in the United Kingdom with a preponderance for the male gender. It is caused by a mutation in a gene normally passed on in an autosomal dominant fashion within the healthy population. Most commonly these tumours affect the sacrococcygeal region (43-85%) and rarely can present like a pilonidal abscess.
We present a case of a 68 year old female with an apparent pilonidal abscess which was found to be suspicious on incision and drainage. An invasive sacrococcygeal chordoma was diagnosed and resected. This case is one that highlights the importance of having a low index of clinical suspicion in older patients presenting with an apparent pilonidal abscess.

Background
Chordoma is a rare, slow growing, malignant primary bone tumour, arising from notochordal remnants. The condition has an incidence of 1 per million per year within the United Kingdom and has an average survival of seven to nine years after diagnosis. It is typically diagnosed in patients between the ages of 40 and 70 years old, with two thirds of cases affecting men.
The responsible gene has been identified as the T-gene, vital for the embryonic development of the notochord. Its mutation (inherited as autosomal dominant) causes uncontrollable cell division and local invasion.
The sacrococcygeal area is the most common site of localisation; with a propensity for local recurrence (43%-85%) with distant metastases being less frequent (20%-40%). Prognosis depends on the quality of resection margins at primary surgery but, is limited by an often incorrect diagnosis preoperatively.
This report highlights a case of sacrococcygeal chordoma, incorrectly diagnosed as pilonidal abscess, leading to unnecessary surgery and potentially delayed referral to specialist services.

Case Presentation
A 68 year old female was admitted to the Northern General Hospital, Sheffield, United Kingdom (UK) with a painful swelling in the natal cleft of six to eight weeks duration, which had acutely worsened in the four days prior to her attendance. She had no symptoms of paralysis, paraesthesia or difficulty controlling her bladder or bowels.
At examination there was a tender, erythematous swelling to the right of the natal cleft. The clinical diagnosis a pilonidal abscess was made and the patient booked for theatre to undergo incision and drainage. No imaging was performed.
At surgery, the “abscess” was noted to drain amorphous material with a disproportionate amount of bleeding. Samples were sent for histology and the wound was packed. Further investigations including Computer Tomography (CT) (figure 1) and Magnetic Resonance Imaging (MRI) scans were requested (figures 2/3). These, in addition to the histology, confirmed the diagnosis of a locally invasive sacrococcygeal chordoma. Upon neurological examination performed post operatively, increased tone with reduced power and brisk reflexes was noted. The patient was referred to the tertiary referral Centre for definitive management.

**Figure 1**– CT scan showing wound packing in place

**Figure 2**– Coronal MRI showing destruction of local structures

**Figure 3**– Sagittal MRI showing extension of Chordoma

**Discussion**

Chordoma is a rare subtype of sarcoma, affecting the spinal column, which can cause local damage to surrounding structures by invasion and compression. It has an incidence of less than 1 in 1,000,000 in the United States and Europe\(^1\). Most patients present between the ages of 40 and 70 and the condition has a preponderance for the male gender\(^2\).

During embryological development, the notochord, a cartilage like structure, acts as primitive scaffolding, supporting the development of the spine. Within
normal development, these remnants persist during adult life. Rarely the T-
gene, inherited in its autosomal dominant form, causes cells of the notochord
remnant to undergo a malignant transformation resulting in this rare type of sarcoma

Commonly, patients diagnosed with sacrococcygeal chordoma present with a
mass in their lumbar or sacral region as tumours need to be quite large to
invade into local structures, causing further symptoms. Patients with more
advanced local disease can present with disturbances of the bladder and/or
bowel function and paraesthesia, pain or paralysis of the lower limbs. 20 to
40% of cases of spinal chordoma metastasize, with common locations
including the lung, bone, soft tissue, lymph nodes, liver, and skin.

Imaging is vital to assess the local and distant spread of the malignancy. The
use of standard CT scan as well as F-18 fluorodeoxyglucose (FDG) positron
emission tomography (PET) scan have been used to identify the subtle
differences between chordoma and benign notochordal cell tumour (BNCT). Subtle
differences within these images can be seen with BNCT showing
osteosclerotic but not osteolytic changes and true chordomas showing
osteolytic changes as well as the presence of a soft tissue lesion. Other types of imaging
techniques have been found to be useful when diagnosing chordoma. On Tc-99m MDP
(740 MBq Tc99m methylene diphosphonate) scan, chordomas usually present with “cold spots” which is
said to be due to increased lytic activity within the lesion, however the images
can rarely show increased uptake. This is due to increased mitotic activity
increasing the vascularity of the lesion.

Histological analysis of tissue is required to make a diagnosis of chordoma.
Open biopsies are the gold standard of diagnosis; however they have increased
risk of causing local damage to structures and therefore are used less
frequency in areas where the lesion may be entwined with delicate structures
such as the spinal cord, or in areas where the lesion is deeper and technically
more difficult to access. In these areas percutaneous needle biopsy could be
used, with lower rates of complication; however this method does have higher
rates failure to obtain a sample.

The management of sacrococcygeal chordoma represents a challenge due to
high rate of reoccurrence. There are a number of treatments options for
chordoma described. Surgery remains the gold standard. Wide local excision
is associated with improved survival and lower rates of recurrence, compared
to marginal or intralesional excision. A combined anteroanterior-posterior
surgical approach to lesions allows suitably wide margins to be achieved. High dose proton radiotherapy has shown promise where primary resection
isn’t possible or in the poor surgical candidate. Improved 5 year survival with
minimal levels of neurotoxicity has been described.
Novel agents such as imatinib, a tyrosine kinase inhibitor, have been used\textsuperscript{13}. This has been shown to stabilise tumour size in 72\% within one cohort study\textsuperscript{14}.

**Conclusion**

This case highlights the importance to maintain a high index of suspicion in patients over the age of 50 presenting with their first “pilonidal abscess”. Thorough clinical examination and appropriate imaging is essential to achieve complete surgical resection and may result in improved outcome.

**References**


Figure 1. CT scan showing wound packing in place
| Figure 2, Coronal MRI showing destruction of local structures |
Figure 3. Sagittal MRI showing extension of chordoma