A chordoma which occurs as a primary tumour outside the axial skeleton is known as an extra-axial chordoma, parachordoma or chordoma periphericum. It is extremely rare and therefore survival, recurrence and the rates of metastasis are not known. Whilst few recurrences have been described, the extra-axial chordoma has the potential for late recurrence at up to 12 years. Metastases are even less frequent. We report the case of a 56-year-old woman who developed an extra-axial chordoma of the right thoracic wall in close relationship with the tenth rib. The tumour was completely removed and the prognosis is excellent.

A chordoma is a rare malignant tumour, which develops from remnants of the embryonic notochord.\(^1,2\) Exceptionally, a tumour with an essentially identical histological appearance to a chordoma may arise outside the axial skeleton. These very rare tumours are referred to as extra-axial chordoma, parachordoma or chordoma periphericum.\(^3-6\) We describe a patient with an extra-axial chordoma in the chest wall.

Case report
A 56-year-old woman presented with a soft-tissue mass involving the right posterior thoracic wall. CT imaging revealed a solid lesion in the soft tissues around the tenth rib, involving bone. A CT-guided needle biopsy identified a tumour which was histologically most consistent with a chordoma or chondroid chondrosarcoma (Fig. 1). Because a definitive histological diagnosis could not be made she was referred to our hospital, where, at open biopsy, a specimen of tumour tissue 2 cm x 2 cm x 1.5 cm was obtained. It had variable cellularity with small groups and cords of vacuolated cells in a myxoid stroma. The cells varied in size and generally had well-defined membranes and large amounts of cytoplasm, ranging from deeply eosinophilic to vacuolated. Many cells had multiple vacuoles, sometimes indenting the nucleus resulting in a lipoblast-like appearance (physalipherous cells). The nuclei were slightly irregular and mitoses were scarce. Immunohistochemically, the tumour cells were positive for epithelial membrane antigen (EMA) and S100 protein. Expression of cytokeratin (CK)-19 and vimentine was seen, and collagen IV and CAM 5.2 (CKs 8 and 18) stains were only weakly positive (all antibodies and immunohistochemistry reagents, DAKO, Glostrup, Denmark). The staining was performed in an automated immunostainer, using a polymer-based system (DAKO Envision, Glostrup, Denmark). A diagnosis of extra-axial chordoma was established. The diagnosis was endorsed by the Dutch National Bone Tumour Group.

Resection of the tumour, including portions of the ninth, tenth and eleventh ribs, and reconstruction of the chest wall using allograft (Composix; BARD Medical, Covington, Georgia) was undertaken. She made an uncomplicated recovery and was discharged on the fifth post-operative day. Figures 2 and 3 show the macro-
Fig. 2
Macroscopic image of the tumour (T). Glistening grey tumour tissue in the soft tissues around the rib and secondarily involving the bony matrix of the rib (R).

Fig. 3
Microscopic image (haematoxylin & eosin staining); cells resembling lipoblasts with vacuolated cytoplasm and centrally located indented nuclei (physaliform) combined with cells with copious eosinophilic cytoplasm set in myxoid stroma.

Fig. 4a
Microscopic images of the tumour (immunohistochemistry). Figure 4a – Pancytokeratin staining (AE1/AE3) was positive throughout the tumour, however only small groups or single cells were stained, many cells did not stain. Figure 4b – The cytokeratin (CK)-10 stain was strongly positive in all tumour cells. Figure 4c – The epithelial membrane antigen (EMA) staining pattern was identical to the CK-10 stain. Figure 4d – S100 staining was only focally positive (cytoplasmic).
scopic and microscopic appearances (haematoxylin & eosin staining) of the resected specimen. Immunohistochemical analysis showed strong positivity for EMA, CK-10 and only slight positivity for S100 protein. Pancytokeratine, CK-7, 34Be12, CK-19, CAM5.2, CK-20, carcino-embryonic antigen (CEA) and calcitriene stains were negative (Fig. 4). The tumour had been completely removed with a 2 mm margin. The prognosis was therefore, thought to be good and no further treatment was deemed necessary. She is being followed up regularly. When seen four months post-operatively she remained disease-free. A CT scan showed no sign of disease.

Discussion
The notochord is an axial mesodermal tissue element, which is present in several different embryonic stages and regresses with advancing maturity. Typically, it is a rod-shaped mass of vacuolated cells, which lies immediately below the spinal cord and may provide mechanical strength to the embryo. Chordomas can occur in several different parts of the body where the notochord has regressed, such as the sacrococcygeal (49% of cases), cranial, sphenooccipital, nasopharyngeal (36%) and vertebral (15%) regions. There is a great diversity of symptoms in the different locations, but all have localised pain as a common mode of presentation. The tumour is not difficult to diagnose, but, because it grows slowly, the onset of symptoms is often insidious, leading to a delay in making the diagnosis. Chordomas are locally destructive and rarely have lymphatic and/or haematogenous metastases. Contrast, local recurrence is frequently seen. Primary treatment should involve radical resection with a wide margin, and this has been identified as the most important prognostic factor for overall and disease-free survival. Unfortunately, definitive radical surgery is often not possible when the tumour is very close to, or involves, vital structures. Radiotherapy is effective but alone will not achieve a cure. Therefore, a combination of surgery and radiotherapy is the standard treatment.

An extra-axial chordoma is macroscopically and histologically similar to an axial chordoma. The main difference is one of location. However, extra-axial lesions are always located near tendinous or bony structures. The differential immunohistochemical staining profiles of axial and extra-axial chordomas are not universally accepted. This is not surprising, considering the immunohistochemical heterogeneity seen within chordomas and any attempt to compare axial and extra-axial chordomas immunohistochemically will lead to some discrepancies. A small series of five patients with extra-axial chordomas was described by Laskowski in 1955. Dabska, in 1977, was the first to use the term parachordoma for tumours mimicking conventional chordoma, but arising outside the central axis. Extra-axial chordoma is more difficult to diagnose than conventional chordoma and it may be difficult to distinguish an extra-axial chordoma from a metastasis or a conventional chordoma. The differential diagnosis includes skeletal and extra-skeletal chordosarcoma.

In this case, the needle biopsy was not contributory to the diagnosis. Tissue obtained by needle biopsy is less likely to give the diagnosis for tumours that have complex architecture and cell pleomorphism. The preferred treatment for patients with extra-axial chordoma is radical surgery. Survival, recurrence and rates of metastasis are unknown because of the rarity of this condition. However, it is known that the tumour may recur after many years and that it seldom metastasises. Six of 37 cases (16%) described in the literature have occurred between three months and 12 years after excision. Two cases (5%) of metastasised extra-axial chordoma have been described. Nevertheless, in most patients long-term disease-free and overall survival may be achieved by radical surgery alone. The role of pre-, peri- and post-operative radiotherapy and chemotherapy is unclear and therefore these modalities should not be advised in the treatment schedule of patients with completely resected extra-axial chordomas. Radiotherapy may be considered in cases where microscopy suggests that the margin of resection is in doubt.

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References