Unusual Cases

SACROCOCCYGEAL TERATOMA IN THE ADULT

A CASE REPORT

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Large sacrococcygeal teratomata are extremely uncommon in adults. The case presented describes a 15-lb. tumour in an intelligent adult female city-dweller who managed to adapt herself remarkably well to its presence.

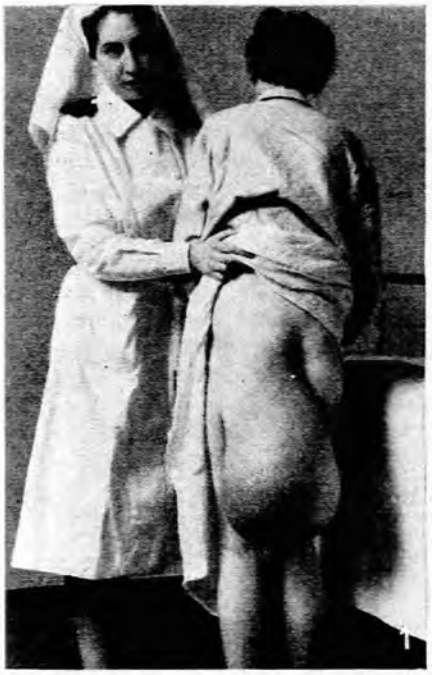
CASE REPORT

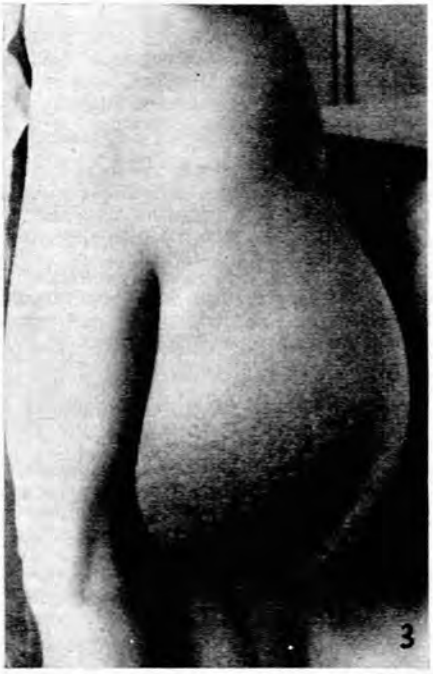
A European female, aged 36 years, was admitted to the Conradie Hospital, Pinelands, Cape, on 2 August 1959 with a large tumour growing from the left buttock. She said that the tumour had been present from birth, and in childhood her parents had consulted 2 doctors about it. It had been decided to leave it alone. The only reason that had brought her to seek medical advice was that 2 days before admission she had felt ill, with shivering attacks, and had noticed an alteration in the appearance of the tumour.

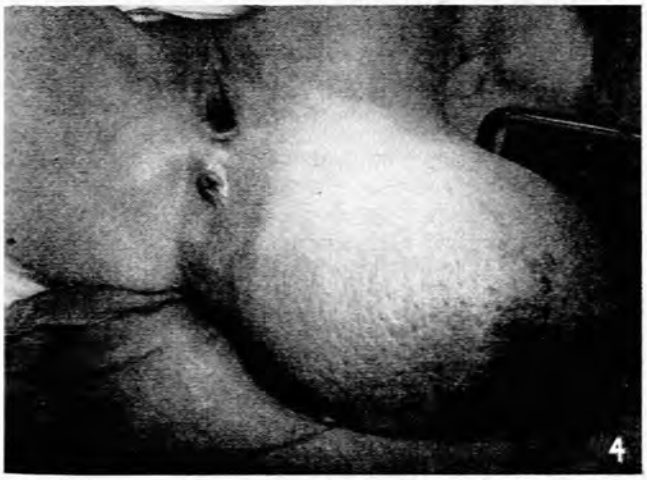
Examination

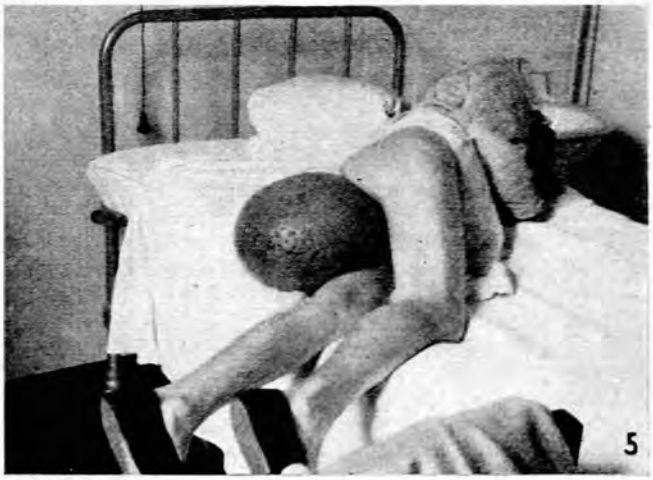
On examination, the patient was an intelligent woman with a slight but well-proportioned physique.

Hanging from the left buttock in the region of the left ischiorectal fossa was an enormous tumour, as seen in the photographs, obviously coeval with the patient. The pedicle,









Figs. 1 - 5. Various views of the tumour.

which extended into the ischiorectal fossa, was in immediate proximity to the vagina and rectum and in fact distorted the anal orifice on the left side. The tumour was firm in consistency, and the skin over the major part had a *peau d'orange* appearance, with a dusky red colour.

The patient was pyrexial with a temperature of 102°F., and it was obvious that cellulitis of the skin over the tumour accounted for her general condition and the changes observed in the tumour. For this antibiotics were given. A straight X-ray picture of the pelvis and the tumour revealed no abnormality.

Operation

On 7 August 1959, under general anaesthesia, the tumour was removed. The patient was placed in the lithotomy position with the tumour resting on a separate table. The dissection was started through a circular incision of the pedicle. Many layers were encountered, one of which was recognizable as a stretched sheet of levator-ani muscle. Eventually the thin wall of an obvious cyst was identified. This cyst was found to fill the left ischiorectal fossa, in contact with the rectum and vagina, and in the final dissection was detached from the last piece of the sacrum and the coccyx. The operation took 3½ hours.

Specimen

This, essentially, was a large skin-covered cyst weighing 15 lb. in all. It contained free fluid and large amorphous masses of sebaceous material, as well as thousands of discrete 'sago-rice' grains of the same material. There was no evidence of other structures such as bones, teeth, hair, etc.

Pathological Report

'The cyst covering was composed of a dome of skin which was 8 inches in diameter. The centre of this dome of skin showed moderate excoriation. The cavity was uniloculated and about 8 inches in internal diameter with contents which were of a soft cheesy consistency and yellow-brown in colour. The cyst lining was generally smooth. The cyst wall was of white, leathery texture and about ¼ inch in thickness.

'The tissue subjacent to the skin consisted of subcutaneous loose connective tissue and layers of muscle into which were infiltrating bands of material resembling the contents of the cyst cavity.

'On microscopic examination, the contents of the cyst showed amorphous material resembling the contents of a sebaceous cyst.

'The lining consisted of avascular fibrous tissue with areas of chronic granulomatous inflammatory reaction and areas lined by cuboidal and ciliated columnar epithelium. One region showed an aggregate of muscle bundles.

'The subcutaneous tissue showed infiltrations of the material contained in the cyst with a chronic inflammatory reaction consisting of foam cells and giant cells.

'The histological features of the growth are those of a cystic teratoma.'

Postoperative Course

Convalescence was normal, and the wound healed by first intention. The function of the sphincter ani was undisturbed. It is interesting to note that it took some weeks for the patient to become accustomed to the absence of the tumour, especially on sitting down.

COMMENTS

Willis,[1] in his series of 82 cases, found the following distribution of teratomata: ovary, 50 cases; testis, 19 cases; and miscellaneous, 13 cases. The 13 miscellaneous cases were distributed at the following sites: epididymis, retroperitoneal space, anterior mediastinum, intrapericardial area, brain, and the presacral area.

Those in the presacral area are commonly referred to in the literature as sacrococcygeal teratomata, and by some authors as Middeldorpf tumours. The propriety of the latter terminology is disputed by others[4] who categorically state that Middeldorpf described neither a presacral teratoma nor any type of sacral neoplasm.

Sacrococcygeal teratomata are well-recognized entities and there is an abundance of case reports concerning them in the literature. One article[5] mentions 426 cases collected from the literature.

Briefly, these tumours are usually found in female infants and are generally obvious at birth. Many of these infants are either stillborn[2] or survive only a few days. The tumours may be solid, cystic, or mixed, and vary in size from that of a walnut to that of the baby's head. These tumours have a definite tendency to become malignant. This tendency has been expressed in different articles as 9%,[2] 17%[4] and 27%.[5] Further, there appears to be a definite association with the incidence of twins.[4,5] One paper[5] reports on a questionnaire sent to patients. Of 23 replies received, 12 confirmed the presence of twins in the family — an association of more than 50%.

Histologically, every kind of tissue, derived from the three germinal layers, has been reported in these tumours, and in some reports portions of limbs and organs are mentioned. The teratomatous cyst, such as the one described here, is a common type and may be confused with the inclusion dermoid cyst. Willis[1] discusses this matter thoroughly, and states that if the cyst wall is examined histologically, evidence of tissue of endodermal and mesodermal origin will be found, as in this case.

The origin of these tumours is a matter of great interest to the theorist, and the presacral area is a happy hunting ground for him on account of its embryological associations.

It is universally agreed that treatment by surgical removal offers excellent prospects of success. It is recommended that the coccyx[5] be removed with the tumour. The prognosis[5] is said to be better with the newborn than with the older child on account of the increased possibility of a malignant change.

DISCUSSION

The great interest of this case lies in the age of the patient and the enormous size of the tumour.

The vast majority of the case references in the literature are of infants and children. In one review of 72 cases,[2] 6 were between 4 and 18 years of age, and a further 6 between 20 and 60 years of age, but no details were given. In another report[6] the patient was aged 22 years, but the cyst involved was a very small one.

It requires little imagination to realize what a handicap a tumour of this size in this particular location could be in every-day life. Sitting down, the choosing of clothes to wear, going to the lavatory, keeping the tumour clean, etc., would be daily problems. Yet this patient had adapted herself so well that, except for an inability to take part in sport, she had led a normal life. She holds a responsible post as secretary, entailing a daily railway journey. She is married and had one pregnancy 12 years ago with a normal delivery of a healthy baby.

The patient, obviously an intelligent woman, states that the possibility of removal of the tumour never entered her head. She blames her parents, particularly her mother, who at no time ever discussed the tumour and its possible significance.

When she married, her husband did not mention the tumour or ask about it. As far as she knows, he is ignorant of its existence. The husband's attitude is odd, because even now he has never asked her for what

operation she has been to hospital. The midwife who confined her 12 years ago did not suggest that she should consult a doctor about the tumour.

In fact, there seems to have been a conspiracy of silence about this tumour by those closest to her. It would appear that she was brought up with the idea that the tumour would be an inevitable part of her existence for the rest of her life. As nobody had ever suggested otherwise, she had become resigned to its presence.

She is delighted to be rid of the tumour, particularly now that she can wear normal clothes and go swimming. She states that she is so adapted to her present state that she can hardly believe she ever had such a tumour.

SUMMARY

1. A case is reported of a 15-lb sacrococcygeal teratoma, dependent from the left buttock, in a 36-year-old European female. The tumour was successfully removed by operation.

2. The physical and psychical difficulties encountered and overcome by the patient while the tumour was present are discussed.

3. The pathology of sacrococcygeal teratomata is briefly discussed.

I wish to thank Drs. J. J. Hofmeyr, E. B. Lawton and H. S. M. van Jaarsveldt for their valuable assistance. Dr. Hofmeyr took the excellent photographs and Dr. R. Shapera prepared the pathological report. I must thank Dr. T. Jones, Medical Superintendent, Conradie Hospital, for access to the hospital records.

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