

Disgerminoma: A case report

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INTRODUCTION

DISGERMINOMA IS AN interesting though uncommon tumour of the ovary. The prefix 'dis' means two, and refers to the fact that it occurs in both sexes. The disgerminoma of the ovary is in fact the same tumour as the seminoma of the testes. Disgerminoma comprises about 3–5% of all malignant ovarian tumours. (Morris & Scully 1958.) It is characteristically a tumour of early life and used to be known previously as 'Carcinoma Puellarium'. A case of disgerminoma showing many characteristic features is described.

CASE REPORT

M.L.F., an unmarried Chinese girl, aged 16 years, was first seen in June 1969. She complained of an abdominal swelling of two months' duration and amenorrhoea for six months. Her menarche was at 14 years, and her periods were scanty, occurring every 3 to 4 months.

On examination, she was a healthy girl, with normal secondary sexominal examination, a smooth firm mass arising from the pelvis up to half way between the symphysis pubis and the umbilicus was present.

She was a virgo intacta and on rectal examination, a mobile mass about 4–6 inches in diameter was felt separate from the uterus. The uterus itself was normal

in size and retroverted.

A pregnancy test (Gravindex) was negative and abdominal X-ray showed no evidence of calcification.

At laparotomy, a soft solid tumour, grey in colour, was found arising from the right ovary. The left ovary was small and had a smooth surface. A right salpingo-oophorectomy was performed, and recovery was uneventful.

Histology

The tumour was composed of pleomorphic cells in solid clumps and, in places, branching cords separated by fibrous strands. There were small follicles of lymphoid tissue separating these cells, features consistent with a diagnosis of disgerminoma.

Follow-up

Since operation, her menstrual period has been occurring monthly and the flow has also been normal.

DISCUSSION

Disgerminomas occur in youth generally before 20 years. It is said to be more common in intersexes and patients having the tumour exhibit signs of varying degrees of hypogonadism. This is similar to the male where imperfectly developed testes are much more

DISGERMINOMA

likely to develop seminoma. However, the tumour itself is not the cause of the sex deficiency which may persist after its removal.

This patient's age — 16 years — corresponds to the classical picture and the scanty and irregular menstruation can be taken as a manifestation of hypogonadism. The surprising thing is that following the removal of the tumour, the menstrual periods have become normal. Although these tumours are most often 'neuter' i.e. not producing any sex hormones, some definitely have mild oestrogenic or androgenic influence. A mild androgenic effect was probably present in this patient, and the removal of this influence could be the reason for the restoration of regular menstruation.

As with many other types of ovarian tumours, the first evidence of its presence is the detection of a mass in the abdomen as in this patient. Although generally there is no characteristic effect on menstruation, as explained above, menstrual abnormalities may co-exist. When marked sex abnormalities are present and an ovarian tumour is detected, the strong possibility of disgerminoma must be considered.

Nevertheless, disgerminoma often occurs in apparently normal women, sometimes first presenting during pregnancy.

Aetiology

The tumour is believed to arise from mesenchymal cells which date back to the early undifferentiated phase of gonadal development. In this phase, the cells have not acquired either male or female characteristics, so that as might be expected the tumour has no effect on the sex characteristics of the patient. Such an origin, as postulated by Meyer, is supported by the fact that an identical tumour occurs in the testicle where it is called seminoma. This is as one would expect with tumours, which as it were lag behind the differentiating process in the gonads, which later develop into either testes or ovaries. Another point in favour of this hypothesis is the fact that in a considerable portion of the reported cases, the tumour occurred in individuals showing some degree of gonadal deficiency.

More recently it has been shown that the tissues of the disgerminoma are **chromatin negative**. There is also evidence that the growth itself or the tissue from which it arises has a sex chromosome complement of XY. (Jeffcoate). According to this view, the disgerminoma arises from 'male' tissue and even in an apparently normal woman, the ovary from which the tumour arises is likely to be the site of mosaicism



Fig. 1. shows the cut section of the tumour.

XX/XY. This mosaicism has been attributed to dispermy, i.e. fertilisation of an ovum by two spermatozoa, one carrying an X and one a Y chromosome. Thus those who develop disgerminoma of the ovary have in their gonads an XY strain of tissue, even though their other tissues have an XX complement only.

Pathology

The disgerminoma tends to grow larger than other sex tumours. In this patient, the tumour was fairly large measuring six inches across. (Fig. 1.) They are solid tumours with a greyish yellow cut surface and they have a characteristic firm and rubbery feel. The growth is usually unilateral though bilateral tumours have been reported.

Microscopically, this is one of the most distinctive and easily recognisable of all ovarian tumours. The

tumour is composed of large cells having a spherical nucleus, which are arranged in bundles or alveoli separated by a network of connective tissue which contain lymphocytes.

Malignancy

According to the majority view, this tumour belongs to the malignant group. There is, however, much variation in the degree of malignancy, and it is extremely difficult to compute its exact incidence. The extreme view is that all are malignant but a figure of 25 – 30% malignancy rate seems reasonable. (Stabler 1963). In many cases, cure has followed simple removal of the tumour. In well encapsulated tumours, the prognosis is good but in the infiltrating variety, the outlook is unfavourable.

Treatment

Surgical excision is the treatment of choice. As these tumours occur in young girls, there is a natural tendency to be conservative and a simple salpingo-oophorectomy is all that is necessary. This surgical conservatism is fully justified by the reported results and by the fact that we are dealing with a tumour of low malignant potential, frequently involving young patients. However, radical surgery may sometimes be indicated if the tumour has infiltrated its capsule and involved adjacent pelvic organs.

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