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Spencer:

Deciduoma malignum

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DECIDUOMA MALIGNUM.

(*With a table of forty cases*)

BY

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ONE of the most interesting diseases which have been observed in recent years is that now generally known by the name of deciduoma malignum, though it has been described under many *aliases*, sarcoma deciduo-cellulare, sarcoma giganto-cellulare, carcinoma syncytiale, sarcoma of the chorionic villi, carcinoma of the chorionic villi, serotinal tumour, &c. The numerous and widely divergent names would seem to indicate either a want of uniformity in the disease or a departure from the ordinary appearances of malignant disease of the uterus, which renders it difficult to explain the origin of the growth, or even to definitely place it in one of the sarcomatous or carcinomatous groups. There are, however, certain features in the recorded cases which are exceedingly interesting and important, and I propose in the following short paper to give an analysis of all the cases published in sufficient detail which I have been able to collect. The total number of cases published up to the present time is, I believe, forty-one, excluding Ahlfeld's tubal case, and cases such as those of Freeborn and Freund, to which no sufficient description is attached. The last published of these forty-one cases (Netzel's) occurred in 1871 and although it appears to have been a well-marked example of the disease under discussion, I have omitted it in order to deal with a round number, and shall limit my analysis to the forty cases contained in the accompanying table. Those who wish to pursue the study of this subject further, will obtain much assistance from the monographs "Sarcoma Deciduo-cellulare" by Pestalozza and by Resinelli in the *Annali di Ostetrecia e Ginecologia* for November, 1895. These two papers contain a full bibliography of deciduoma malignum and its cognate subjects, which has been of great use to me in the preparation of the accompanying table. It would not be difficult for a hostile critic to take exception to the inclusion of some of these forty cases. Thus the last two of Pestalozza's may be objected to as possibly cases of simple hydatid mole; but it seems to me fairer to accept the description of these two by the author of four other cases in which the disease was certainly malignant, than to criticise them without the opportunities of studying them which the author himself enjoyed. This form of criticism *à distance* has been indulged in somewhat freely in the case of the disease we are considering, and it may be generally remarked that the opinions adverse to deciduoma malignum as a distinct pathological entity, proceed entirely from those who have no personal acquaintance with it. In any case there will be found in the table no less than twenty-seven cases in which metastatic deposits existed in other organs, and thus place the malignancy of the disease beyond dispute. Deciduoma malignum may be defined as a malignant disease of the body of the uterus (occurring presumably at the placental site), arising in association with, or more commonly subsequently to, pregnancy, and rapidly terminating fatally, with secondary growths in various organs of the body, more especially the lungs and vagina. It is characterised microscopically by large cells of various shapes, of which some occur singly or in groups, and others in fused masses with large deeply-staining nuclei resembling enormous giant cells, the so-called "*syncytium*." This syncytium when it is at all abundant (in my own cases it formed the principal part of the tumour), gives to the growth a very remarkable appearance under the microscope, totally unlike that of any other growth I have met with in the uterus. It resembles closely the syncytium or layer of fused cells which forms the outer coating of the normal chorionic villus, and it seems probable that the syncytium of deciduoma malignum develops from the syncytium of the chorionic villus, though this is denied by some authorities. As to the origin of the syncytium of the chorionic villus there is still much dispute. While some consider that it arises from the epiblast of the fœtus, others are of opinion that it arises from maternal mesoblast. The deciduomal syncytium is, like the normal syncytium, vacuolated. Another characteristic feature of the growth is the presence of extensive hæmorrhages, which are due to the marked tendency of the malignant growth to eat its way into the blood-vessels. The first author who described the development of malignant disease of the body of the uterus subsequently to delivery was Chiari, who under the title " Ueber drei Fälle von primaren Krebs im Fundus und Corpus des Uterus," published an account of three cases, of which two occurred in young women aged 22 and 23. All three cases terminated fatally within about six months of delivery, and presented similar appearances at the necropsy and under the microscope. Chiari remarks upon the peculiarity that they lead to death a very short time after a labour or abortion. He also alludes to a case examined by Dr. Juergens, who left the diagnosis between sarcoma and carcinoma *in suspenso*, considering it a mixed form of the two, " an appearance possibly brought about by the resemblance to sarcoma cells of decidual cells shortly after an abortion." Chiari thought that the growth in his cases was of the nature of carcinoma, but he has since been convinced that they are examples of deciduoma malignum.

This important publication of Chiari's apparently attracted little attention, although it found its way into Ruge and Veit's classical work on " Cancer of the Uterus," where most gynæcologists must have been struck with the remarkable fact of cancer developing in the body of the uterus at such an early age.

Cases by Tibaldi, Guttenplan, and Meyer followed, but it is only since the publication of Saenger's case in 1889 that widespread interest has been aroused in this new disease. This case was designated by Karg as " malignant metastasising deciduoma," the term deciduoma having previously been employed by other authors. Saenger himself, however, appears to have preferred the term sarcoma deciduo-cellulare, and it appears to me that credit is due to Chiari rather than to Saenger for calling attention to the development of malignant disease of the body of the uterus after pregnancy. Although numerous cases have been observed upon the continent, no example of the disease had been reported in this country until last year, when I made a preliminary report to the Obstetrical Society of a case which I had observed in 1889. The case has recently been read in full at the same society, together with a case by Mr. Morison.

On reference to the table it will be seen that the age of the patients varied between 22 and 55 years.

6	patients	were	between	20	and	25
11	„	„	„	25	„	30
8	„	„	„	30	„	35
4	„	„	„	35	„	40
6	„	„	„	40	„	45
3	„	„	„	45	„	50
1	„	„	„	50	„	60

The number of pregnancies previous to the one with which the disease was associated is given in the fourth column of the table. The figures may not be in every instance quite accurate, since it is not always clear whether the last pregnancy is included or not; but from the fourth and fifth columns combined we get the following facts:—The patients had all been pregnant at least once before the disease developed, the history being doubtful in one case only (Jeannel's); the disease appears to be almost entirely confined to multiparæ, and in some cases there had been a long interval between the last two pregnancies. *In* 18 (45 *per cent.*) *of the cases there is a history of hydatidiform mole.* Usually the symptoms arose within a few weeks of the labour or abortion; in one case a secondary growth was observed 8 days, in another 15 days after delivery, showing that the disease in these cases existed during pregnancy. In some instances no symptoms of the growth occurred for several months after delivery.

The characteristic symptoms presented by the disease are hæmorrhages coming on within a few weeks after labour or abortion, the passing of masses of growth, extreme anæmia, fœtid discharge and septicæmia with or without rigors. In a few in-

TABLE OF FORTY CASES OF DECIDUOMA MALIGNUM.

Author.	Reference.	Age of Patient.	No. of Pregnancies.	Relation of symptoms to Labour or Abortion.	Hysterectomy.	Result.	Growths found in.
Chiari (I.)	*Wiener Med. Jahrb.*, vii., 1877, p. 364	24	?	Repeated hge. and discharge followed labour at term	No	Death (6 mths.)	Irregular inner surface uterus; small circumscribed nodules in wall of ut.; also in both broad ligaments.
Chiari (II.)	,, ,, ,, ,,	23	2	Hge. 4 wks. after third child (at term); hæmoptysis	No	Death (6 mths.)	Irregular masses in mucous membrane of fundus and upper part of body of ut.; in lungs.
Chiari (III.)	,, ,, ,, ,,	42	?	Hge. 6 days after labour (at sixth mth.); hæmoptysis	No	Death (6 mths.)	Irregular mass in ant. wall of ut.; also small circumscribed nodules; also in lungs, ovary, vagina, pelvic lymphatic glands.
Tibaldi	*Gazetta d. Osped.*, 1882, No. 58	31	4	Continuous hge. after last child (fifth)	No	Death	Uterus, brain, lungs, kidney, colon, ovary.
Guttenplan	*Dissert. Inaug. Strassburg*, 1883	28	7	Hge. began 3 mths. after hydatid mole; hæmoptysis	No	Death	Uterus, lungs, vagina.
Meyer	*Archiv f. Gynäk.*, 1888, vol. xxxiii., p. 53	55	3, last 10 yrs. before	Hge. since, hydatid mole passed 6 mths. before	No	Death (9 mths.)	Uterus (designated by Prof. Klebs, "Epithelioma papillare").
Saenger	*Centralbl. f. Gyn.*, 1889, p. 132	23	0 (?)	Hge. for 3 wks. after labour (8 mths. ch.); in fourth week septicæmia; afterwards hge. and discharge ceased	No	Death (7 mths.)	Uterus, iliac fossa, lungs, diaphragm, ribs (designated by Karg, a "malignant metastasising deciduoma").
Pfeifer	*Prag. Med. Wochenschr.*, 1890	35	4, normal (1 ab.)	Hydatid mole, Dec., 1888; hge. in Sept., 1889 (? aborted then)	No	Death (Feb. 4, 1890)	Uterus (fundus and left wall), vagina, lungs.
Pestalozza	*Annali di Ost. e. Gin.*, 1895, p. 743	25	? 1	Abortion, Feb. 1, 1888	No	Death (Aug. 1, 1888)	Uterus, especially anterior part and fundus; vagina, broad ligaments, lungs.
Pestalozza	,, ,, ,, ,, p. 744	33	5, normal	Hge. began shortly (over a month) before death	No	Death	
Pestalozza	,, ,, ,, ,, p. 745	45	9, normal	Hydatid mole, Dec. 26, 1889; irregular hges. afterwards	No	Death (March 30, 1891)	Uterus (upper segment), disseminated nodules ant. and post. walls; lungs.
Pestalozza	,, ,, ,, ,, p. 765	32	7, normal	Hge. after labour, April 17, 1894	No	Death (Oct., 1894)	Uterus and vagina; no p.m.
Pestalozza	,, ,, ,, ,, p. 770	22	2 (1 at term., 1 ab.)	Hge. after abortion (hydatid mole, 2 mths.), Feb., 1894	Vaginal hyst. (May 14, 1894)	Recovered (well after one year)	Uterus.
Pestalozza	,, ,, ,, ,, p. 772	44	12, all normal except first, (ab. from injury)	Hydatid mole, Oct. 4, 1894	Vaginal hyst. (Oct. 13, 1894)	Recovered (well after one year)	Uterus.
Gottschalk	*Archiv f. Gyn.*, vol. xlvi., p. 1	42	2 ch., 3 ab.	Abortion (2 mths.), curettage followed by blood-stained discharge for 6 wks., then hge. recurred	Vaginal hyst. (Aug. 16, 1892)	Death (March 11, 1893)	Uterus, pelvis, lungs, spleen, right kidney.
Schmorl	*Centralbl. f. Gyn.*, 1893, p. 169.	?	?	Hge. 12 wks. after labour	No	Death (6 mths.)	Uterus, lungs.
Koettnitz	*Deutsch. Med. Wochensh.*, 1893, No. 21.	25	2, normal	Normal labour (third), then hge. with slight intervals; fever, rigors, delirium	No	Death (10 wks.)	Uterus, vagina, lungs.
Löhlein	*Centralbl. für Gyn.*, 1893, p. 297	47	7	Hydatid mole, May, 1890; cessation of menses in summer of 1891; then hge. and discharge after Feb., 1892; then for 2 mths. quite well; then fever and discharge	Vaginal hyst. (Aug. 8, 1892)	Recovered (well Jan. 7, 1893)	Uterus.
Fraenkel	*Archiv f. Gyn.*, vol. xlviii., p. 80	25	0	Hydatid mole (third mth), July, 1892; parametritis; then well till Feb., 1894; then hge.	No	Death (June 9, 1894)	Uterus, bladder, parametrium, spleen.
R. Klien	*Archiv f. Gyn.*, vol. xlvii., p. 243	27	2 ch., 1 ab. (Aug., 1892)	Hydatid mole, March 12, 1893; then hge. and pains; curettage, May 15, 1893; fever, rigors, hge.	No	Death (Nov. 11, 1893)	Uterus (body and cervix), vagina, small pelvis, lungs.
P. Müller	*Verh. d. Deutsch. Gesell. f. Gynäk.*, IVten Kongress, 1891, p. 341	30	6	Abortion induced (5 mths.); some wks. later masses felt in ut.; hges.; ut. emptied; then a row of cystic tumours appeared in post vaginal wall	No	Death (5 mths.)	Uterus, vagina, gluteal region.
Menge	*Zeitschr. f. Geb.*, vol. xxx., p. 323	35	8 ch., ab. 1	Hydatid mole, Dec. 28, 1892; hge. and curettage, May, 1893; hge. and curettage; small nodule removed, July, 1893	Vaginal hyst (Aug. 11, 1893)	Death (6 mths. after operation)	Uterus, vagina, vestibule, pelvis, lungs, bronchial glands, spleen, stomach, jejunum, right femur.
Lebensbaum	*Centralbl. f. Gyn.*, 1893, p. 112	27	3	Labour (fourth) at term; hge. 5 wks. later, lasting 11 days; curettage; hge. increased; rigors	Vaginal hyst. (July 3, 1891)	Death (6 days)	Uterus, vaginal wall.
Hartmann and Toupet	*Bull de la Soc. Anat. de Paris*, 1894, p. 723	25	1	Hge. (?) a year after labour	No	Death (8 mths. after first symptoms)	Uterus, p.m. examination incomplete.
Nové Josserand and Lecroix	*Annales de Gyn.*, 1894, p. 100	24	2 ch. (last, 2½ yrs. ago)	Regular after last ch.; pregnant again Dec., 1892; hydatid mole passed; hge. began 1 mth. after passage of mole	Vaginal hyst. (July 12, 1893)	Recovered (well 3 mths. later)	Uterus.
Marchand	*Monatschr. f. Geb.*, 1895, p. 513	34	9 ch. (last, Nov. 26, 1893)	Hge. 3 wks. after last child	Vaginal hyst. (April 20, 1894)	Recovered (well Oct., 1894)	Uterus (fundus).
Schauta	*Centralbl. f. Gyn.*, 1895, p. 248	29	4	Hge. 1 month after hydatid mole (fifth pregnancy)	Vaginal hyst. (Nov. (?) 21, 1894)	Recovered (well 1½ year later)	Uterus, vagina (post wall).
Superno	*Annali di Ost. e. Gin.*, 1895, p. 775	32	5 (5th ch. 10 mths. before)	3 mths. hydatid mole; hge. and pain	Vaginal hyst. (Sept. 14, 1892)	Recovered (well 1 year later)	Uterus.
Resinelli	,, ,, ,, ,, p. 813	28	3, normal (last, March 28, 1890)	Hge. and discharge followed abortion (3 mths.) in Nov., 1891	No	Death (March 14, 1892)	Uterus, left tube, vaginal wall, lungs, liver.
Jeannel	*Archives de Tocol.*, 1895	26	?	? Abortion in Jan., 1893, then regular till March, 1894	Vaginal hyst. (May 3, 1894)	Recovered (well Dec. 20, 1894)	Uterus.
Kuppenheim	*Centralbl. f. Gyn.*, 1895, p. 916	33	5	Hge. 3 wks. after last ch. (June, 1894)	Vaginal hyst. (Aug. 20, 1894)	Recovered (well June, 1895)	Uterus.
Tannen	*Archiv f. Gyn.*, 1895, p. 94	23	2	Hydatid mole (third pregnancy), July, 1893; then regular and well till Jan., 1894, when hge. began	Vaginal hyst. (June 30, 1894)	Recovered (well 9 mths. later)	Uterus.
W. Williams	*Amer. J. of Obs.*, 1895, p. 319; *Gynæcological Trans.*, 1895	35	5	Labour, April 15, 1894 (ch. dead), post part hge.; on 15th day of puerperium nodule in right labium majus; septicæmia	No	Death (July 12, 1894)	Uterus (fundus and posterior wall), labium, lungs, spleen, liver, kidney, right ovary.
Bacon	*Amer. J. of Obs.*, 1895 p. 679	48	8 ch., ab. 2	Hydatid mole (9th mth.), Dec., 1892; hge. 5 wks. later, and every 2 wks. till June, 1893	No	Death (June 25, 1893)	Uterus, right broad ligament, lungs.
Runge	*Archiv f. Gyn.*, 1896, p. 185	44	Multip. (last ch. 3 yrs. ago)	Rather profuse hge. for 5 mths.; hydatid mole passed	Abdominal hyst. (Oct. 28, 1895)	Recovered (well 3 mths. later)	Uterus.
Apfelstedt and Aschoff	*Archiv f. Gyn.*, 1896, p. 511	33	2 ch.	Abortion (4 mths.), Oct. 4, 1894, fœtus macerated; Profuse menstruation, Dec. and Jan., with bloody discharge between; a "polypus" removed; discharged Feb., re-admitted May, with bloody discharge	Vaginal hyst. (May 24, 1895)	Death (June 19, 1895)	Uterus (body), to within a few m.m. of peritoneum, lungs, stomach, pancreas.
Apfelstedt and Aschoff	*Archiv f. Gyn.*, 1896, p. 511	42	2 (1882, 1886)	Hydatid mole, March 28, 1895; 8 *days* later painful swelling in left labium majus; on incising this metastatic hydatid mole found	No	Death (July 25, 1895)	Uterus, paravaginal cellular tissue, l. labium, lungs, spleen.
Lonnberg and Mannheimer	*Centralbl. f. Gyn.*, 1896, p. 474	42	2, normal	Hydatid mole (4 mths.), Nov. 19, 1895; hge. 7 wks. later	Abdominal hyst. (Oct. 18, 1895)	Recovered (well April 1, 1896)	Uterus and vagina (vaginal metastatic growths removed by operation, Oct. 30, 1895.)
Morison	*Obst. Trans.*, 1896	35	9 ch.	Hge. after labour 9 weeks before	Vaginal hyst. (Dec. 11, 1894)	Death (July 11, 1895)	Uterus.
Spencer	*Obst. Trans.*, 1896 (preliminary, 1895)	27	1 ch. (7 yrs. before)	Masses of growth passed, and hge. 3 wks. after normal labour and puerperium (ch. living)	No	Death (within 10 wks. of labour)	Uterus (body and cervix), lungs.

stances, but not usually, hæmoptysis has been observed, due to the secondary growths in the lungs; in others secondary nodules having somewhat the appearance of varicose veins have appeared in the vagina or labium. *In one case* (Apfelstedt and Aschoff's) *the secondary growth in the labium and paravaginal tissue consisted of a hydatidiform mole*, with the usual stalks and cysts; in another (Müller's) the metastatic vaginal growths took the form of a row of cystic tumours. In several cases delirium was observed; my patient died from thrombosis of the cerebral veins, probably septic in origin. In 16 cases the labour occurred at or about term, at least later than the sixth month of pregnancy. In 6 cases abortion occurred in the earlier months. The disease is rapidly fatal unless treated by hysterectomy, which has been performed in 17 cases, of which 15 were by the vaginal and 2 by the abdominal route. Of these 17 cases only 2 died from the immediate effects of the operation (six and twenty-six days after); 3 others died about six months after the operation, and 12 remained well when reported at various intervals up to eighteen months after the operation. The two most interesting of the cases submitted to hysterectomy are those of Schauta, and of Lonnberg and Mannheimer, as in both these cases secondary growths were also removed from the vagina, and yet the first of these patients remained well after a lapse of a year and a half (see *Monatschrift für Geb. und Gynäk.*, 1896, p. 387), and the second patient after half a year.

Metastatic deposits were observed in the lungs in 19, or nearly half the cases. The next most frequent seat is the vagina, where they were met with in 13 instances; next in frequency comes the spleen, 5 cases; the broad ligament, 4 cases; the ovary, kidney, vulva and pelvis, of each 3 cases; the stomach, intestines, liver and glands, of each 2 cases; the brain, diaphragm, ribs, iliac fossa, bladder, femur, Fallopian tube, pancreas and gluteal region, of each 1 case.

Such are the facts known about this interesting disease; but they have not been allowed to prevail against the theories of the critics who have directed their attention to trying to prove that the disease is nothing more than an ordinary sarcoma of the uterus unconnected with pregnancy (the association being a mere coincidence). If we merely confine our attention to the histology of the growth, we must, of course, admit that neither the large size of the individual cells nor the plasmodial masses (the so-called syncytium) is necessarily an indication that the growths have originated in the products of conception, though it is possible that in some cases the extensive development, the insular arrangement, the vacuolation, or the peculiar staining of the syncytium may prove to be different from the plasmodia of ordinary sarcoma. But, so far as I know, in spite of the interest which the disease has aroused in the last six years, not one of the critics has been able to produce a single case of sarcoma of the uterus exhibiting the syncytium and occurring in a woman in whom pregnancy could be excluded. This piece of evidence must be forthcoming if the criticism which regards deciduoma malignum as an ordinary sarcoma is to have any weight. The histological elements are remarkable chiefly for the large size of the cells and for the syncytium. This syncytium closely resembles the syncytium met with as the outer covering of the chorionic villus and the budding and hypertrophied syncytium of the hydatidiform mole (as described by Fraenkel and others). One is, therefore, naturally led to look to the normal syncytium for the origin of the abnormal growth, a view which is rendered the more attractive by the publication of Apfelstedt and Aschoff's case, in which the secondary growth was a hydatidiform mole, an observation which, if corroborated, proves at once the origin of the growth from the chorionic villus.

The facts which, in my opinion, go to show that the disease is *not* an ordinary sarcoma are (1) the constant association with pregnancy (there is, I believe, no proof of its preceding or occurring apart from pregnancy); (2) the association with hydatidiform mole in at least 45 per cent. of observed cases, but possibly in a much larger percentage, since early hydatidiform disease is not always obvious at the first glance; (3) the constant occurrence of the disease in the body of the uterus; (4) Apfelstedt and Aschoff's case already cited, and perhaps Müller's case; (5) the macroscopic appearance of the growth, which has been mistaken for placenta by obstetricians of large experience; (6) the microscopic appearances.

For these reasons I regard deciduoma malignum as a distinct pathological entity, and as a disease arising from the products of conception. Whether it arises from the maternal or from fœtal structures, whether it should be called a sarcoma or a carcinoma, is comparatively unimportant, and it may be left to future research to decide. "Deciduoma malignum" may be as bad a name for the disease as its critics assert, but the object of a name is to serve as a label and not necessarily as a definition, and it fulfils that purpose sufficiently well.

In the six years of its existence this little pathological bantling has suffered at the hands of its critics no more grievous injury than this, that, not content with denying its legitimacy, they strive to deprive it of its maternal name.