2.0 Gy given with 5 fractions/week. After a cumulated dose of 26 Gy there was a two-week pause partly due to service of the treatment machine. The second series was given with our ordinary three-field technique up to 68 Gy corresponding to a CRE-value of 1 880 reu. The acute radiation side-effects were quite similar to those observed in otherwise healthy individuals.

Four weeks after conclusion of radiation therapy no tumor could be palpated and the acute radiation side-effects had receded. After three months the patient was still in an excellent clinical condition, although his CD4 count was as low as 0.0006×10^9/l. The patient will be controlled at regular intervals at the oncological department with regard to his cancer, whereas the problems associated with his AIDS will be handled at the department of infectious diseases.

The therapeutic strategy chosen seems to have been satisfactory for this patient, although one cannot exclude the possibility that the further deterioration of his CD4 count was accelerated by the radiation therapy.

Key words: AIDS, anal carcinoma, case report.

C. SVENSSON
M. KAIGAS
E. LIDBRINK
S. GOLDMAN

Department of General Oncology and Department of General Surgery
Södersjukhuset
Stockholm
Sweden

April 1991

Corresponding author: Dr Christer Svensson, Department of General Oncology, Södersjukhuset, S-118 83 Stockholm, Sweden.

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INCOMPLETE CURRARINO SYNDROME WITH A PRESACRAL LEIOMYOSARCOMA

Currarino syndrome is a very rare condition characterized by the triad anorectal malformation, sacral bony abnormality and a presacral mass. The sacrum defect was first recognized in 1837 by Bryant (1). The entire triad was first described by Kennedy in 1926 (2), and later on defined as a unique complex of anomalies by Currarino et al. in 1981 (3). Autosomal-dominant hereditary transmission has been found in about 50% of the patients (4). The presacral mass has been reported to be a malignant tumour in four cases (5-8). All were malignant teratomas. We here report on a woman with an incomplete Currarino triad and a presacral leiomyosarcoma.

Case report. A 59-year-old woman was in January 1991 submitted to postoperative radiation therapy at our hospital after surgery for a presacral leiomyosarcoma. The tumour size was $13 \times 11 \times 7$ cm. The pelvis was irradiated from anterior and posterior fields up to a total midpoint dose of 50 Gy in 25 fractions in 5 weeks (CRE-value 1557). Since birth her defecation had been painful and the diameter of the faeces had usually been less than 1.5 cm. Three years before diagnosis a grandchild of the patient was born with a classical Currarino syndrome. The daughter and mother of the patient have always suffered from constipation problems. A brother of the patient died 2 years old of meningitis. Due to the family history the patient was examined for a possible Currarino syndrome. Rectoscopy revealed a stenosis of the rectum 13 cm from the anal ring. Roentgenography of pelvis showed no defects in sacrum or coccyx. A CT-scan of the pelvic region and abdomen did not show any residual tumour.

Discussion. The Currarino syndrome is a triad of anorectal stenosis, sacral anomaly and a presacral mass. In members of the same family, one or two basic features of the syndrome may be lacking, suggesting an incomplete form of the syndrome (3). The patient had no sacral bony defect. Eighty per cent of the patients are diagnozed by 16 years of age. The range in age is from 1 day to 65 years (3). The most frequent symptoms are lifelong constipation, recurrent perianal infections and urinary tract problems secondary to neurologic impairment or the extrinsic mass (5, 9, 10).

The embryogenesis of the syndrome was first postulated by Currarino et al. (3). The endodermal layer (future gastrointestinal tract) is normally in close approximation to the neural ectoderm (future neural tube) in early fetal life. The notochord and somities eventually fuse to form the vertebral body and separate the gastrointestinal tract from the spinal chord. If the notochord is split or displaced laterally, the anterior fusion of the vertebral body will fail. This allows a fistula to be formed between the gastrointestinal tract and the spinal channel with potential enteric elements ventrally and neural elements dorsally. Partial resorption of the fistula may give rise to a meningocele or an enteric cyst. The anorectal anomaly is a consequence of these early abnormal adhesions between the hind gut and the neural tube.

The presacral teratoma is formed by local enteric and neuroectodermal elements combined with mesodermal elements brought into the space by the developing somities and other local mesodermal structures. The presacral mass is usually a meningocele, teratoma, hamartoma, enteric duplication or a combination (9, 11). Kirks et al. (9) found that the presacral mass was a meningocele in 48% and a teratoma in 39% of the patients. Only four cases of malignant presacral tumour have been reported (5-8). In our patient the leiomyosarcoma probably developed from an innate presacral mass. She had had symptoms of a presacral tumour and rectal stenosis since birth. Her leiomyosarcoma was attached to the sacrum and not to the uterus or the ovaries. Aschcraft & Holder (5) believed that a benign presacral tumour could undergo malignant changes. Heij et al. (11) expressed the opinion that a patient with a known presacral benign teratoma without symptoms might be followed by rectal examination and serum alpha-fetoprotein determinations.

Two patients with Currarino syndrome and meningocele who initially presented with meningitis have been reported (10, 12). There is thus a possibility that the patient's brother, who died of meningitis 2 years old, also suffered from a Currarino syndrome.

The rectal stenosis can be caused by external compression of the presacral mass or by adhesions. Extension of the teratoma into the rectal wall has also been reported (5, 6, 13). The stenosis is usually located distally (11), while our patient had her stenosis somewhat higher up in the rectum.

The sacral bony defect varies from lateral deviation of the coccyx to unilateral absence of the lower sacral segments (5). Our patient had no sacral bony defect.

When a patient presents with a family history of a lifelong constipation, the Currarino syndrome should be kept in mind. The examination for this syndrome should include rectal examination, roentgenography of sacrum with contrast in the rectum and a CT-scan of the pelvis (4, 11, 14). If available a magnetic resonance imaging (MRI) will be of help to detect anomalies of the spinal channel (11).

In most cases conservative treatment with laxatives or repeated anorectal dilatations will not be satisfactory. The results of excisional surgery are good (11). Removal of the tumour by a posterior approach was most frequently chosen by Aschcraft & Holder (5). The patients were placed in jack-knife position with an obturator in rectum. The coccyx and the presacral tumour were excised and if necessary also the attached rectal wall. Our patient had been operated on with an anterior approach due to size and localization of the tumour.

Key words: Currarino syndrome, presacral leiomyosarcoma.

J. NORUM
E. WIST
L. BOSTAD

Department of Oncology
and Pathology
University Hospital of Tromsø
Tromsø
Norway

April 1991

Corresponding author: Dr Jan Norum, Department of Oncology, University Hospital of Tromsø, N-9012 Tromsø, Norway.

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