Primary bone sarcomas of the pelvis represent the most challenging problem in limb-saving surgery. Chondrosarcoma is the most common type, followed by osteosarcoma and Ewing’s sarcoma. The symptoms are often vague, so that the diagnosis is made late, when the tumor has already reached a large size. Nowadays hindquarter amputation is reserved for complex tumors involving the sacrum, the sciatic nerve, the iliac vessels or the bladder. Internal hemipelvectomy should be undertaken whenever possible; the reconstructive techniques are based on the three anatomic regions concerned: ilium, periacetabular area, and pubis. Complications occur in more than 50% of the patients. The 5-year survival rate is about 55%. Special scores, such as the MSTS/ISOLS and the TESS score, should be used to make the various studies comparable.

Keywords: pelvis; bone sarcoma; surgery; limb-saving.

INTRODUCTION

The most frequent bone sarcomas of the pelvis are chondrosarcoma, osteosarcoma, and Ewing’s sarcoma; however, pelvic bone sarcomas constitute less than 10% of all malignant bone tumors (23). The overall incidence of malignant primary tumors of bone is 1/100,000/year (3), so that pelvic primary bone sarcomas account for an incidence of less than 1/1,000,000/year. The thin cortical bone and the large content of the pelvis result in delayed diagnosis, so that the tumors often reach a huge volume, which explains why their prognosis is worse than for similar tumors in the appendicular skeleton.

Management of bone sarcomas, including the biopsy, should be restricted to specialized centers. Nevertheless, the average clinician must be aware of the immense threat these tumors create: all too often a periacetabular chondrosarcoma is mistaken for osteoarthritis of the hip.

CHONDROSARCOMA

Chondrosarcoma most frequently involves the pelvis (16). The tumor may be very large, but often low-grade. It contains a myxoid substance that can lead to seeding via the biopsy track or via contamination of the operative field. Microscopic examination alone can be insufficient to distinguish a low-grade chondrosarcoma from a benign chondroma: clinical and radiographic data should also be taken into consideration. Chemotherapy and radiation therapy have only limited value; they are mostly used for recurrences or unresectable tumors.

OSTEOSARCOMA

The pelvis is an uncommon site of origin of osteosarcomas, with fewer than 10% of all such lesions there (16). For primary osteosarcoma, the
peak incidence is in the second and third decade of life. The high frequency of bone-to-bone spread (especially to the sacrum), the high prevalence of vascular involvement and the fact that there are no major fascial anatomical barriers in the pelvis to prevent extension explain the poor outcome. Of the treatment options that are currently available, adjuvant chemotherapy pre- and postoperatively seems to be indicated, since the ability to achieve control of the tumor with operative treatment alone is limited. Radiation therapy can be helpful as palliative treatment.

**EWING’S SARCOMA**

About one fifth of all Ewing’s sarcomas are situated in the pelvis (16). It is the second most common location. In most cases they are voluminous tumors, and they have a high propensity to metastasize, especially to the lungs.

The tumor initially arises within the medullary canal and destroys the cortex while causing periosteal elevation, leaving the so-called Codman’s triangle. After high-dose polychemotherapy and local radiation local recurrences are known to develop in a high percentage of cases. Therefore Ewing’s sarcomas probably need additional surgical resection, although the gain was not statistically significant in several studies (11, 17, 25).

**CLINICAL ASPECTS AND DIAGNOSIS**

Sarcomas in the pelvis and in the acetabular area in particular are frequently discovered because of pain produced in the hip joint on weight bearing. Also nonspecific complaints, such as tiredness and loss of weight should be taken seriously. An abdominal mass, complaints about fullness, obstruction and bladder symptoms are also possible (16).

Plain x-rays reveal the presence of the tumor lesion; most often it appears as a capricious lytic mass, though standard x-rays alone are insufficient to make the diagnosis. Computer tomography (CT scan) is very effective for defining the extent of the tumor, both within the bone and in the soft tissues surrounding it. Magnetic resonance (MRI) is invaluable in the planning of limb-sparing surgery. It is ideal for revealing the marrow, which enables one to evaluate the intramedullary extension as well as the extraosseous component of the tumor; this has proven especially helpful in detecting small lesions and skip metastases. Bone scintigraphy helps to determine polyostotic involvement, the existence of metastases and the intraosseous tumor extension.

**STAGING**

Besides the previously described procedures, the work-up consists of thoracic and abdominal CT-scan and laboratory analyses including plasma electrophoresis. If necessary arteriography, as well as a contrast study of the bowel and intravenous pyelography, will give further evidence of encroachment of the mass on the organs in the pelvis.

All these investigations, together with the result of the biopsy will determine the surgical stage: IA: low-grade, intracompartmental; IB: low-grade, extracompartmental; IIA: high-grade, intracompartmental; IIB: high-grade, extracompartmental and III: metastases (10).

**BIOPSY**

The biopsy technique is more important in the pelvis and the acetabular area than anywhere else. The type of biopsy used will vary according to the radiographic presentation of the tumor. When a very definite diagnosis can be made radiographically, frequently a confirmatory needle biopsy can be done. When chondrosarcoma is highly suspected, an open or closed biopsy may be done. This should be planned so that the biopsy area can be effectively sealed off from the resection to prevent contamination. The surgeon who will be responsible for the excision of the tumor should do the biopsy.

**CLASSICAL HEMIPELVECTOMY OR HINDQUARTER AMPUTATION**

Hemipelvectomy has been the conventional procedure for surgically treatable primary malignant
bone tumors of the innominate bone. Unfortunately, this operation is not only mutilating but it also sacrifices a normal leg. On the other hand, partial or total internal hemipelvectomy preserves the lower extremity and can be as effective as conventional hemipelvectomy in eradication of the tumor. Nowadays hindquarter amputation is only performed if internal hemipelvectomy is impossible because of involvement of crucial anatomical structures. In case of a posteriorly located tumor a large anterior myocutaneous flap with the superficial femoral artery and vein can be developed as an alternative (26).

INTERNAL HEMIPELVECTOMY OR LIMB SAVING SURGERY

With this technique one resects the entire hemipelvis or a part of it, preserving a functional intact distal extremity. In case of partial hemipelvectomy one may distinguish three major types of resection, according to Enneking. Type I resections are those of the iliac wing, not including the acetabulum, type II resections include the acetabular region (with or without the hip joint) and type III are the ischiopubic resections.

Eilber et al. (7) were among the first to use this technique in the late Seventies. Enneking and Dunham (8) described the utilitarian incision that provides access to both the intra- and extrapelvic aspects of the innominate bone. This incision begins at the posterior inferior iliac spine, follows the crest of the ilium and the inguinal ligament as far as the femoral vessels where it turns distally along the rectus femoris for 5 to 7.6 centimeters and finally curves laterally to end just posterior to the shaft of the femur at the junction of the proximal and middle thirds of the thigh.

In the original article of Enneking and Dunham (8) the reconstruction was kept simple. After iliac excision (type I) the remaining part of the ilium was brought up to, and when feasible fused with the sacrum. After periacetabular resection (type II) the femoral head was denuded of its articular cartilage and fixed to the remaining ilium, in order to obtain a pseudoarthrosis or a fusion. However, when the hip joint was to be removed en bloc, the remaining part of the femur was fixed to the stump of the ischium or pubis, also to create a pseudoarthrosis or a fusion. After resection of the pubis (type III) only the soft tissues of the pelvic floor were reconstructed.

O’Connor and Sim (22) described various alternatives such as the use of intercalary allografts. They mentioned that only 50% of the attempted fusions succeeded and that infection was the most frequent complication (23%). Flail limbs and pseudoarthroses had less good function than fusions.

Later on, more sophisticated techniques were developed. Harrington (15) excised the pelvic tumor en bloc in four patients, autoclaved the resected part of the pelvis, reimplanted it, and used a cemented total hip prosthesis as a supplement. Only one patient had a graft-related problem after 8.5 years, more specifically a fracture.

Grimer et al. (14) excised, irradiated and reimplanted four pelvic tumors: one became infected and required removal of the graft; two patients died of metastases and the fourth was well after 2.5 years.

Mnaymneh et al. (20) replaced the whole pelvis, except the pubic bone, with a massive allograft in one patient with a chondrosarcoma. The patient kept his own femoral head. This patient did well; he was asymptomatic for 5.5 years in spite of a partial resorption of the iliac crest, a hairline fracture of the graft and removal of lung metastases.

Thompson and Manivel (27) implanted a whole-joint allograft in a single patient, but neuropathic arthropathy developed, and the necrotic head had to be replaced with a total hip prosthesis.

Langlais and Vielpeau (19) used hemipelvic allografts in combination with a total hip prosthesis in four patients. They had no infections or dislocations, but one patient sustained a fracture of the acetabulum after 17 months. Their early results after a mean follow-up of only 19 months were good. Harrington used similar hemipelvic allografts in ten patients. After an average of five years, there were graft-related problems in two of them: recurrent dislocation in one and fatigue fracture in both. In one of the fatigue fractures the replacement allograft became infected, but this could be
controlled by debridement. The results were mostly good to excellent.

Aboulafia et al. (1) used a Nieder saddle prosthesis in 17 patients. It consists of a femoral stem and a V-shaped upper end, designed to articulate with the remaining ilium. They noted five prosthesis-related complications, especially instability of the prosthesis.

Tumor centers in Birmingham and Stanmore (2) used a custom-made cemented acetabular prosthesis in combination with a total hip prosthesis and this without closing the pelvic ring distally. After an average follow-up of seven years, they found 23% prosthesis-related problems in 35 patients: recurrent dislocation in 17%, femoral loosening in 3% and polyethylene wear also in 3%. Moreover they noted a 26% infection rate and local recurrences in 24%. Their total complication rate was about 60%, which was confirmed by others, including Campanacci and Capanna (4). However, it is noteworthy that hindquarter amputation also leads to a 35% complication rate, with perioperative death in 9% (5).

Mutschler et al. (21) used a (sub-) total hemipelvic polyacetale prosthesis in nine patients, where a total hip prosthesis could be cemented into the artificial acetabulum. Their average follow-up was 52 months. They had major complications in seven cases of which three were prosthesis-related. Gradinger et al. (13) proposed an adaptable subtotal metal prosthesis. Six out of nine patients had major complications, but only one was prosthesis-related. Their average follow-up period was 27 months. It is too early to evaluate such a (sub-) total prosthesis, which completes the pelvic ring.

COMPLICATIONS OF INTERNAL HEMIPELVECTOMY

Campanacci and Capanna (4) reviewed 105 patients after various kinds of internal hemipelvectomy, and noted complications in 56%. The complication rate was equally distributed among the three anatomical regions according to Enneking. The infection rate was 29%, vascular and visceral problems occurred in 15%, and the same was true for mechanical problems.

Abdominal hernia was present in 12%; nerve damage was noted in 10% and cardiopulmonary problems in 3%.

PROGNOSTIC FACTORS

A study from the Memorial Sloan-Kettering Cancer Center in New York, where they surgically treated 103 pelvic sarcomas (18), showed a local recurrence rate of 34%. It was significantly correlated with tumor size and surgical margins. Metastases, mostly to the lungs, occurred in 35%. They were significantly correlated with the surgical stage and with hindquarter amputation, but the latter was probably due to the fact that amputation was reserved for the worst cases. The 5-year survival rate was 55% for chondrosarcomas, 52% for Ewing’s sarcomas and 47% for osteosarcomas. It was significantly correlated with size, surgical stage, hindquarter amputation and surgical margins. Interestingly there was a distinct flattening of the survival curve after 4 to 5 years, suggesting a more favorable outcome after this critical period. The New York group also felt that involvement of the sacrum should call for hindquarter amputation.

CONCLUSION

Pelvic sarcoma is one of the most challenging problems in limb-saving surgery. The challenge intensifies when critical structures such as the sacrum, the sciatic nerve, the iliac vessels, the bladder or the peritoneum are involved. In these cases hindquarter amputation should be considered.

In the future standardized systems should be used to report the end results of pelvic resections and other tumor surgery. The MSTS/ISOLS score (9) is now widely accepted. However, according to Davis et al. (6), quality of life should be given more impact when evaluating outcome, and therefore they developed the TESS score.

REFERENCES


**SAMENVATTING**

**J. SOMVILLE, S. VAN BOUWEL. Primaire botsarcomata in het bekken.**

Primaire botsarcomata in het bekken vormen de grootste uitdaging op het gebied van lidmaatsparende chirurgie. Chondrosarcoma komt het meest voor, gevolgd door osteosarcoma en Ewing’s sarcoma. De symptomen zijn dikwijls vaag, zodat de diagnose pas laat tijdig wordt gesteld, wanneer de tumor reeds volumineus is geworden. Tegenwoordig gebeurt een hemipelvectomy van het oude type alleen nog voor tumoren die het sacrum, de n. ischiadicus, de vasa iliaca, of de blaas aantasten.
Zo veel mogelijk moet een inwendige hemipelvectomie worden uitgevoerd; de reconstructie is gebaseerd op de 3 anatomische regio’s van het bekken: ilium, peri-acetabulaire streek, en pubis. Verwikkelingen doen zich voor in meer dan 50% van de gevallen. De overleving na 5 jaar is ongeveer 55%. Speciale evaluatieschalen, zoals de MSTS/ISOLS en de TESS score, dienen gebruikt te worden om de verschillende studies onderling meer vergelijkbaar te maken.

**RÉSUMÉ**

**J. SOMVILLE, S. VAN BOUWEL. La chirurgie des sarcomes primitifs du basin.**

La résection-reconstruction des sarcomes primitifs du basin constitue le plus grand défi possible pour le chirurgien. Le chondrosarcome est le plus fréquent, suivi par l’ostéosarcome et le sarcome d’Ewing. Très souvent les symptômes sont vagues, si bien que le diagnostic n’est établi que tardivement, quand la tumeur est déjà volumineuse. De nos jours, l’hémipelvectomie classique est réservée aux tumeurs complexes avec invasion du sacrum, du nerf sciatique, des vaisseauxiliaques ou de la vessie. Si elle est possible, l’hémipelvectomie interne est préférable. La reconstruction est basée sur les trois régions anatomiques du bassin : l’os iliaque, la région périacétabulaire, et le pubis. Les complications sont fréquentes : elles surviennent dans plus de 50% des cas. La survie à 5 ans est approximativement de 55%. Il est indiqué d’utiliser des échelles spéciales, comme la MSTS/ISOLS et la TESS, pour faciliter la comparaison d’études diverses.