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ORIGINAL STUDY

Manifestations and management of osteochondromas : A retrospective analysis of 382 patients

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Osteochondromas represent the most common primary bone tumours ; they reportedly represent 20-50% of all benign bone tumours and 10-15% of all bone tumours. Malignant transformation is their most severe complication. However, deformities and interference with major joint function are the most frequent complaints in patients with hereditary multiple osteochondroma. Treatment should therefore aim not only at surgical resection of the masses but also at prevention of deformities.

This article reports observations made on 69 patients with hereditary multiple osteochondroma and 313 patients with solitary osteochondroma, with a mean follow-up of 13.4 years.

Keywords : hereditary multiple osteochondroma ; osteochondroma ; malignant transformation ; bone deformity.

INTRODUCTION

Osteochondromas represent the most common bone tumours and are developmental lesions rather than true neoplasms (*3, 13*). They can be present at birth and continue to appear and grow throughout childhood and into puberty (*10, 13*). They can be solitary or multiple (*10, 13*). Treatment of patients with Hereditary Multiple Osteochondroma (HMO) is much more problematic and complex than that of patients with solitary osteochondromas (*24*). They appear in different clinical presentations depending on the joints affected and the number of osteochondromas (*20, 24, 25*). Therefore surgical treatment is often directed towards correction of the associated deformities rather than restricted to the osteochondromas alone (*2, 5, 13, 14, 24*). The treatment sometimes continues throughout the entire life, which makes the situation troublesome both for the surgeon and the patients. Repetitive operations and progressive deformities may cause excessive impairment for the daily living of the patients (*24, 25*). Malignant transformation is also a severe complication (*1, 26*). Surgeons should therefore consider reducing the number of surgical sessions in these patients.

This study deals with 69 patients with HMO and 313 with solitary osteochondromas and aims to describe clinical presentation and evolution, and attempts to provide clues on treatment planning.

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Table I. — Patient's demographics

	Solitary	HMO
Number of patients	313	69
Age	21.1 (8-77)	21.3 (3-69)
Gender (male/female)	204/109	49/20
Number of surgeries		
1	296	9
2-5	17	23
6-10	–	32
11-20	–	4
> 20	–	1
Malignant transformation	7 (2.2%)	8 (11.6%)

PATIENTS AND METHODS

Between 1986 and 2003, 423 patients with osteochondromas were given surgical treatment in the Department of Orthopaedics and Traumatology of Ankara University, Faculty of Medicine. Patients with inaccessible and/or insufficient files (n = 9), and those with follow-up less than 24 months (n = 32) were not included in the study. The remaining 382 patients had enough follow-up and their data were investigated retrospectively with the permission of the institutional review board (table I).

Regional radiographic films and skeletal surveys were available for 313 patients with solitary osteochondroma and 69 patients with HMO, and CT scans for 92, MRI studies for 37, bone scintigraphy for 14, and angiographic evaluation for six patients (fig 1). Osteochondromas were resected marginally and the clinical diagnoses were confirmed by histopathological methods postoperatively. The files were re-evaluated and clinical profiles including heredity, descriptions, clinical presentation, follow-up period, were defined for each patient. The mean follow-up period was 13.4 years (between 2 and 19 years). Non-parametric tests were used for statistical analyses.

RESULTS

Solitary osteochondromas

Solitary osteochondromas were present in 313 patients. The mean age at the first admission to our hospital was 21.1 years (range, 8 to 77) and male to female ratio was 204/109. The main complaint was a palpable mass (91% of the cases). Pain was present in 35% of cases, usually related to the soft tissue disturbance by the bone mass and complications such as bursa formation (over the greater trochanter in 1 case), osseous deformity and/or movement limitation of a major joint – ankle (n = 2), knee (n = 4), shoulder (n = 2) –, and nerve compression (carpal tunnel syndrome in 1 case, peroneal nerve palsy in 1 case). Most of the lesions (84%) were found at the distal femur (fig 2), proximal tibia, proximal humerus, scapula, proximal femur, distal radius, ilium, vertebrae, talus, distal tibia, and distal humerus (fig 3). The remaining 50 osteochondromas (16%) were located on the proximal fibula (6), ischium (6), calcaneus (6), distal ulna (6), ribs (6), metacarpal bones (5), metatarsal bones (5), hand phalanges (5), foot phalanges (3), and clavicle (2). Most of the lesions (81.2%) were in the vicinity of major joints : the knee (39.9%), the shoulder (20.5%), the wrist (6.7%), the ankle (6.4%), the hip (5.1%), and the elbow (2.6%).

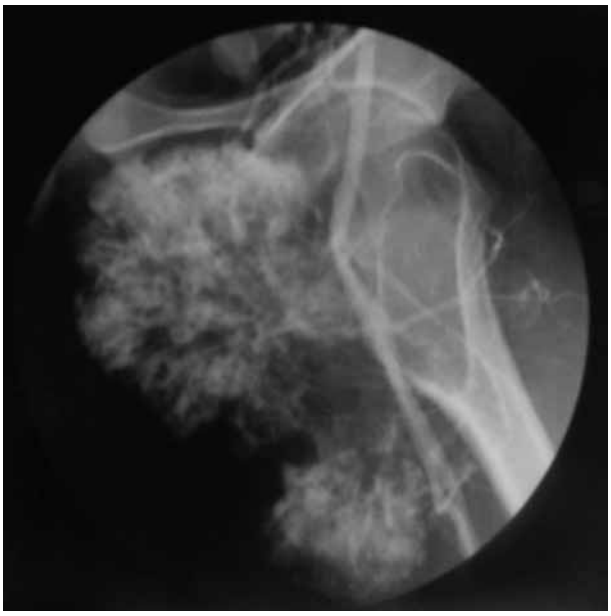

Fig. 1. — Secondary chondrosarcoma, Grade 1, in a female aged 29 years : A huge tumour mass is seen adjacent to the femoral artery (Case 11).

The incidence of sarcomatous degeneration was 2.2% (7 patients) in this group. Localisations were proximal femur (2), distal femur (2), proximal tibia (1), proximal fibula (1), and proximal humerus (1). The mean age at the time of diagnosis

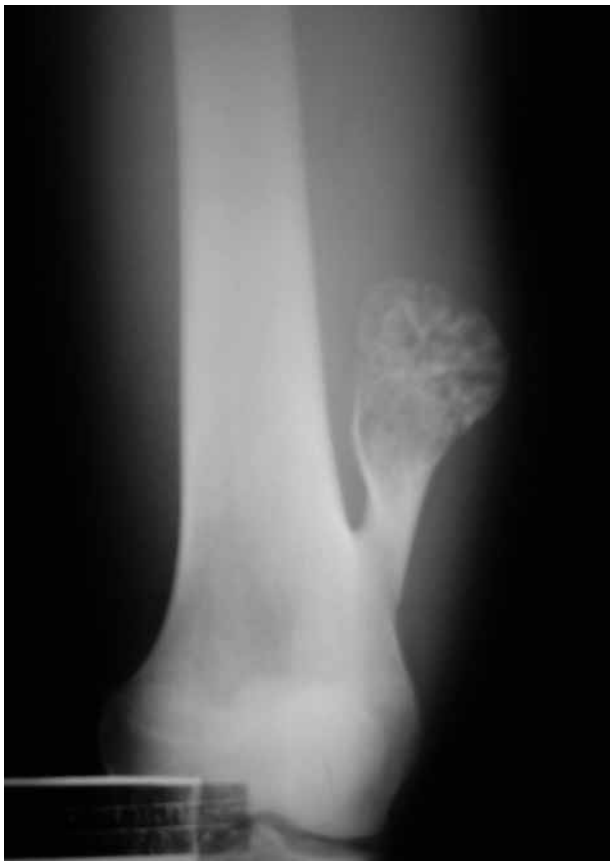

Fig. 2. — Pedunculated osteochondroma on the distal femoral metaphysis in a male aged 19 years.

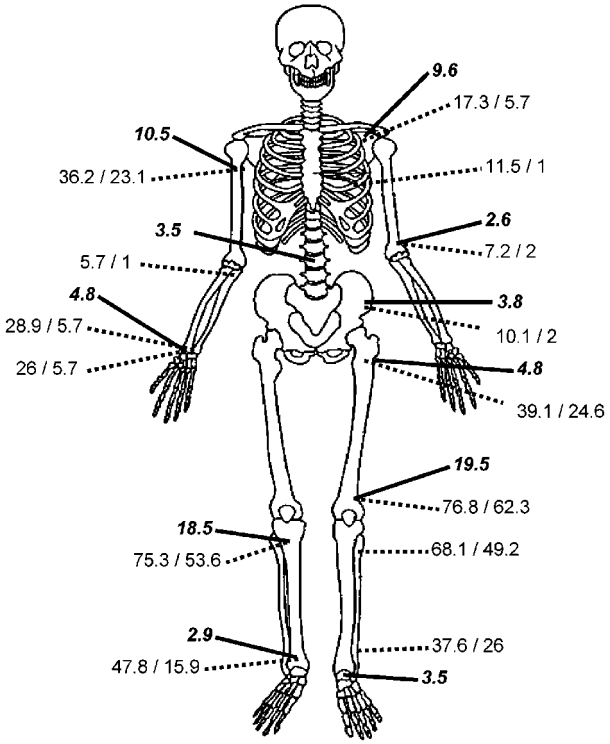

Fig. 3. — Distribution of the lesions : Bold lines represent the percentage (%) of lesions of the patients with solitary osteochondromas and dotted lines represent the percentage (%)/ bilaterality ratio (%) of lesions of the patients with HMO.

in patients with malignant transformation was 29.6 years (range, 18 to 63).

Hereditary Multiple Osteochondroma

HMO was seen in 69 patients. The mean age at the first admission was 21.3 years (range, 3 to 69) and the male to female ratio was 49/20. The main complaints were a palpable mass (100%), pain (87%), deformity (81%), joint impairment (78%), and short stature (42%). There were several specific complaints such as bursitis (7), and nerve palsy (2). Among the patients with multiple osteochondroma, 42 (60.8%) were referred to our institution from other centers after their initial diagnosis and/or treatment. There was no significant difference regarding age on first admission between the HMO and solitary osteochondroma groups (Mann Whitney U, p = 0.587). The condition was established to be familial in 43 patients (62.3%) and not familial in 26 (37.7%). The average number of lesions was 14 per individual (range : 6 to 37). The lesions were rarely seen on vertebrae (3), talus (3), clavicle (2), navicular bone (1), proximal radius (1), pubis (1) (fig 3). A combination of distal femoral, proximal tibial and proximal fibular osteochondromas was seen in 37 cases (53.6%), bilaterally in 30 cases (43.4%). Distal tibial and distal fibular lesions were also present in 24 cases (34.7%) and 2/3 of them were bilateral. Distal radial and distal ulnar lesions were also present in 12 cases (17.3%) and one-third of them were bilateral. No proximal ulnar and proximal radial lesions were found together on the same joint. Approximately 78% of our patients had a clinically recognisable joint impairment, most

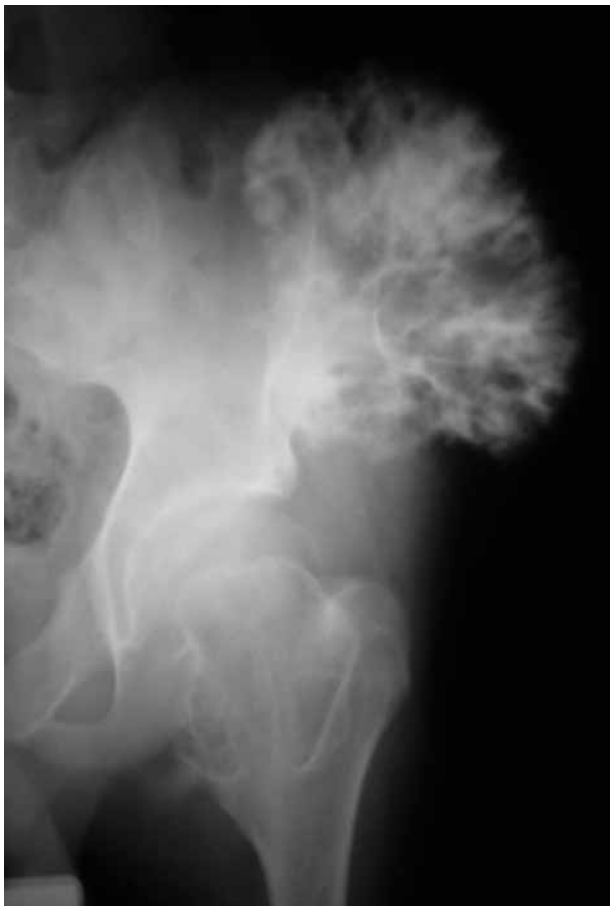

Fig. 4. — Secondary chondrosarcoma, Grade 1, in a male aged 36 years : The patient had no evidence of disease six years after marginal resection (Case 12).

commonly involving the forearm (63%), the ankle (42%), or the knee (17%). Short stature below the fifth percentile was the main problem for both male and female patients (39% and 48% respectively). Patients underwent multiple surgical procedures at different sites, 5.6 times on average (ranging from 1 to 21). Seventeen patients (24.6%) needed corrective osteotomies. The incidence of sarcomatous degeneration was 11.6% (8/69) for the HMO group. Localisations were the iliac bone (2), proximal femur (2), distal femur (1), proximal tibia (1), scapula (1), and one patient had sarcomatous transformation simultaneously in the scapula, ilium and distal femur (fig 4). The mean age of those patients was 36.2 years (range, 15 to 69).

The size was measured for 701 excised lesions. The dimensions varied between $1.5 \times 0.5 \times 1$ to $29 \times 7.5 \times 12.5$ centimeters. Cartilage cap thickness was measured for 385 benign lesions between 4 to 16 mm. Local recurrences of osteochondromas were seen from 1 to 3 times in 21 patients (6.9%) with solitary osteochondroma. There were between 1 and 4 local recurrences in 16 patients (23.2%) with HMO. The average interval between surgery and the first local recurrence was 24 months (range : 11 to 45) for patients with solitary osteochondroma and 17 months (range : 7 to 81) for patients with HMO.

Malignant transformation was seen overall in 15 cases (table II). Of those, 12 were Grade 1, three were Grade 2. There was no Grade 3 tumour. Ten of them had a previous histopathological diagnosis of osteochondroma. The remaining five (three with HMO and two with solitary osteochondroma) patients were initially operated on, with the suspicion of a secondary chondrosarcoma over osteochondroma at the first admission. Two patients, who had marginal excisions, had local recurrence but there were no recurrences after wide/radical resections. One patient who had Grade 2 secondary chondrosarcoma needed scapulothoracic disarticulation two years after the initial scapulectomy because of recurrence. Chemotherapy and radiotherapy were given to one patient (Case 8) and he died after five years follow-up period from multiple metastases. Chemotherapy only was given to another patient (Case 15) who was considered to be inoperable, due to end-stage malignancy, and he died three months after the diagnosis. There was no significant difference between the ages at diagnosis of chondrosarcomatous transformation in the HMO and solitary osteochondroma groups (Mann-Whitney U, $p = 0.463$).

DISCUSSION

Osteochondroma has been reported to represent 20-50% of all benign bone tumours and 10-15% of all bone tumours (*13*). Its clinical and radiological features are often pathognomonic, making diagnosis easy (*3, 19, 25*). From 1986 to 2003, the number

Table II. — Demographics of patients with malignant transformation

No	Type	Age	Sex	Localization	Surgery	Gr	Nux	Met.	F-U (year)	Status
1	S	18	M	Prox. humerus	Wide excision	1			15	NED
2	S	35	F	Prox. tibia	Wide excision	1			7	NED
3	S	30	F	Prox. femur	Marginal excision Hip disart.	2	+		7	NED
4	S	27	M	Prox. femur	Hip disart.	1			6	NED
5	S	26	M	Distal femur	Wide excision	1			5	NED
6	S	39	F	Distal femur	Wide excision	1			4	NED
7	S	32	M	Prox. fibula	Prox. Fibula resection	1			4	NED
8	HMO	31	M	Scapula	Scapulectomy Scapulotoracic disart	2	+	+	5	Exitus
9	HMO	15	M	Scapula Ilium Distal femur	Wide excision	1			8	AWD
10	HMO	32	M	Distal femur	Marginal excision High femoral amp.	1	+		15	NED
11	HMO	29	F	Prox. femur	Marginal excision	1			9	NED
12	HMO	36	M	Ilium	Marginal excision	1			6	NED
13	HMO	69	M	Prox. femur	Hip disart.	1			5	NED
14	HMO	29	F	Prox. tibia	Wide excision	1			3	NED
15	HMO	49	M	Ilium	Inoperable	2		+	-	Exitus

(No : Number of patients, Gr : Grade, Met : Metastases, F-U : Follow-up, S : Solitary, HMO : Hereditary Multiple Osteochondroma, M : Male, F : Female, NED : No evidence of disease, Prox : Proximal, Disart : Disarticulation, Exc : Excision, Res : resection, Amp : Amputation, AWD : Alive with disease).

of benign bone tumours diagnosed and/or treated in our institution was 1,454 and osteochondroma patients constituded 29% of them. Osteochondromas are either sessile or pedunculated, and they vary widely in size and number (*3, 13, 25*). Carroll *et al* (*6*) found that the percentage of sessile osteochondromas correlated with the extent of deformities with more severe angular deformity when more than 90% of osteochondromas were sessile. Most of the lesions were sessile in our HMO patients (76%). Both pedunculated and sessile lesions were seen in 13%. Pedunculated lesions were seen in 64.8% of patients with solitary osteochondroma. Osteochondromas are usually located in the metaphyseal region of the proximal humerus, distal radius and ulna, proximal tibia, and distal femur, but can also be found on the pelvis and scapula, calcaneus, metacarpals, navicular bone and ribs (*13, 24, 25*). All lesions affecting long bones in our patients were in the metaphyseal regions, predominantly at the knee, hip and shoulder. In our patients with solitary osteochondroma, the lesions affecting long bones of the lower extremity were most frequently symptomatic ; long bones in the upper extremity were less frequently affected (in 24.9% versus 50%). As with other bone tumours, osteochondromas occurred most often about the knee (39.9% of the cases). The femur was the single bone most frequently affected (24.3% of the cases) with distal involvement being four times more common than proximal involvement. Tibial osteochondromas accounted for 22% of cases and most commonly occurred in a proximal location. The humerus was also a frequent site for osteochondromas (11% of the cases). Other more unusual locations of osteochondroma included small

bones of the hands and feet (8.3% of the cases), and pelvis (6%). These findings are in line with literature data (*13*).

HMO is an autosomal dominant inherited trait with an incidence of at least one in 50,000, which makes it one of the most common inherited musculoskeletal conditions (*7, 12, 13, 20*). Most HMO patients are reportedly diagnosed by the age of 5 years and virtually all have been diagnosed by 12 years (*10, 13*). The mean age of our patients was 21.1 at the first admission. This divergence with the literature might come from the fact that 42 (60.8%) of our patients had been diagnosed and /or treated in other centers before referral to our clinic. Peterson (*18*) and Malagon (*12*) respectively stated that 65% and 64% of HMO patients had a family history. This ratio was 59.6% in our series.

The number of osteochondromas, the skeletal distribution, the degree and type of angular deformity, and even the rate of malignant transformation vary significantly in HMO, even within families (*13, 24*). Clinical presentation may be either bilateral or not (*13, 24, 25*). They can cause multiple and severe complications including pain, restricted range of joint movement, deformities and shortening of the long bones, and nerve or blood vessel compression (*4, 13, 16, 21, 23, 25*). Carroll *et al* (*6*) noted that the amount of involvement and deformity of the forearm and distal leg were a measure of the overall disease extent. Increased number of osteochondromas, prominent angular deformity and shortening in these areas indicated more severe, generalised involvement. The distribution was mostly bilateral in our patients. Bilaterality was obvious at the knee (81.1%), shoulder (64%), hip (64%), ankle (47.6%), and wrist (21%).

The cartilage cap measurement is important for osteochondromas (*1, 9, 13, 22, 26*). In a study by Hudson *et al* (*9*), the thickness of the cartilage cap in benign osteochondromas averaged 9 mm, with a maximum of 2.5 cm. The mean cartilage thickness in our patients was 6.6 mm. For lesions with malignant transformation, mean measurements of 3.9 cm (between 0.5 and 15) and 4.6 cm (between 1 and 15) were reported by Ahmed *et al* (*1*) and Wuisman *et al* (*26*), respectively. In our study, the cartilage cap thickness was irregular on the lesions with malignant transformation, with a maximum measurement of 11 cm.

The most severe complication is malignant transformation of an osteochondroma (*1, 13, 26*). The suspicion of secondary chondrosarcoma is indicated by growth of the tumour after puberty, the presence of pain, or a thickness of the cartilaginous cap over 1 cm, extensive calcifications, and irregularities within the cartilage mantle, erosions or destructions of the adjacent bones, in adults (*13, 19, 25*). Pain was present in 12 (80%) of our patients with malignant transformation and growth of the mass after puberty was obvious in 10 (66%). The exact incidence of malignant transformation of osteochondroma is not known, especially for solitary lesions, as a number of these are asymptomatic and are never identified (*13, 19*). According to the literature, the incidence is between 0.4% and 2% in patients with solitary osteochondroma and between 1 and 4% in patients with HMO (*1, 13*). The reported risk of malignant transformation of osteochondromas in patients with HMO is up to 27.3% in Garrison's *et al* (*8*) reports. In more recent analyses, the risk of malignant transformation has been from 0.6% to 2.8% with the lowest figure of 0.57% reported in a French study of 175 HMO patients by Legeai-Mallet (*11*). However we should keep in mind that as the follow-up period gets longer, the incidence of malignant transformation will increase. Thus the incidence of malignant transformation was reported as 7.6% for the solitary osteochondroma group and 36.3% for the HMO group from the Mayo Clinic (*1*). The reason for these high rates may be a referral selection bias but also a longer follow-up period. In our series, the malignant transformation rate was 2.2% in the solitary osteochondroma group and 11.6% in the HMO group ; 12 of the lesions were Grade 1 and the remaining three were Grade 2. Seven patients were cured by wide excision. Five patients needed radical resection. Three of them had recurrent malignancy following previous resections. Even though the rate of local recurrence is reported to be primarily dependent on the adequacy of surgical therapy rather than on histological grade, we believe that localisation and histological grade also influence the survival of the patient (*1, 13, 15*). Especially

there is considerable difficulty to achieve wide resection of intrapelvic tumours (*1, 13*).

No multicentric malignant transformation was reported in the series of the Mayo Clinic in 46 patients (*1*), and Wuisman *et al* (*26*) reported two patients with bifocal chondrosarcomas among 27 patients with HMO. Overall, there are a few reported cases of multicentric chondrosarcomatous transformation in HMO patients (*1, 17*). In our series, one patient with HMO had multifocal malignant transformation (Case 9).

Half of the HMO patients with malignant transformation were reported to have no initial histopathological diagnosis of osteochondroma (*1*). This ratio in our patients was 5/15.

Centrally located osteochondromas about the pelvis, hips and shoulders are reported to be particularly more prone to undergo malignant transformation (*1, 13, 26*). The cause may be the delayed diagnosis of the lesions in these areas (*1, 13, 22*). One of our patients (Case 15) who was considered to be inoperable at the time of initial diagnosis had a giant mass over the ilium which was extending from pelvis to abdominal cavity, invading the sacral roots. Garrison found an even distribution of sarcomatous transformation in solitary osteochondromas between long and flat bones and in 80% a predilection for flat bones in multiple osteochondromas (*8*). In our series in HMO patients in whom malignant transformation occurred, the localisation was the knee in two patients, the hip in two, the ilium in two, the scapula in one, and both scapulae, iliac wing and distal femoral bone altogether in one patient. For solitary osteochondromas, malignant transformation was seen at the knee in four patients, at the hip in two and shoulder in one patient. Overall, nearly half of the malignant transformations occurred in the knee region (7/15). It has been reported that malignant transformation typically occurs at different ages, depending on whether it is associated with solitary osteochondroma (average age : 50-55 years) or HMO (average age : 25-30 years) (*13, 26*). We could not confirm this conclusion in our series : no significant difference was found between patient ages at the time of diagnosis of malignant transformation in the solitary osteochondroma and HMO patients.

Even though osteochondroma is a benign tumour, malignant transformation is one of its severe complications. Patients with solitary or multiple osteochondromas should be warned of the risk of malignant transformation, and regular follow-ups should be emphasised. In addition, deformities and major joint impairments are the most frequent complaints in HMO patients. Improvement in the daily living of the patients and prevention of deformities can be provided by timely scheduled surgery, in the light of the natural history of the lesions.

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