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Regression of a Sessile Osteochondroma: A Case Study and Review of the Literature

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The most common benign bone tumor is osteochondroma. Its natural history is poorly understood as a consequence of its benign symptomatology in most cases. It typically grows in childhood and growth during adulthood can be a sign of malignant degeneration. We present a case of spontaneous regression of a solitary osteochondroma. A review of the current literature which reveals 21 other cases of spontaneous regression of solitary osteochondromas is also discussed. We believe that the possibility of regression of solitary osteochondromas should be taken into account when considering surgical excision.

Introduction

The most common benign bone tumor is osteochondroma, accounting for up to 35% of all benign bone tumors and 8% of all bone tumors [1-4] . It most commonly occurs in the metaphyseal region of the long bones[4,5]. It presents as a bony exostosis with a cartilaginous covering on the external surface of the lesion[5] as a consequence of an osteochondral formation caused by the subperiosteal development of an aberrant island of cartilage evolving by enchondral ossification[5,6]. Depending on the installation characteristics in the primitive bone, two types can be distinguished: sessile osteochondroma, which has a broad base, and pedunculated osteochondroma, which develops from a narrow shaft of bone[5,7].

Most lesions appear in children and adolescents as painless, slow-growing masses and may go unnoticed. Its incidence is probably higher than published. Eighty five percent of bone tumors diagnosed as osteochondromas are solitary lesions. The remaining cases are part of a syndrome known as Multiple Hereditary Exostoses Syndrome (MHE/MHO)[4]. Recently, biallelic inactivation of the EXT1 locus was described in nonhereditary osteochondromas. Reduced EXT1 or EXT2 expression may be associated with defective endochondral ossification which is likely to be involved in the formation of osteochondromas.

Its natural history is poorly understood as a consequence of its benign symptomatology in most cases. It typically grows in childhood, and growth during adulthood can be a sign of malignant degeneration. We present a case of spontaneous regression of a solitary osteochondroma and a review of the current literature. We believe that the possibility of regression of solitary osteochondromas should be taken into account when considering surgical excision.

Case Report

An 11-year-old girl was evaluated for a few week history of a solitary right arm mass and mild tenderness on palpation. The pain was related to physical activity, however, it did not interfere with sport's performance. There was no history of trauma, night pain, fever, sweats or chills. The family history, medical history, social history, and complete review of systems were not contributory. On physical examination, the patient seemed to be a healthy girl with a firm, fixed mass, mildly tender to palpation, not attached to skin or deeper layers of soft tissue, and of bony consistency. The mass was located at the anteromedial aspect of the middle third of her right humerus. No other masses were palpable. Range of motion of the shoulder and elbow were normal. No neurovascular deficit was found on examination.

On plain x-rays (Fig. 1A,B) at the time of presentation, a solitary sessile osteochondroma was identified at the anteromedial aspect of the middle third of the humeral shaft. There was no clinical or radiographic evidence of hereditary multiple exostoses. Treatment options such as operative (excision) versus non-operative (observation) alternatives were discussed with the patient and her family. The family chose to observe the lesion.

At one-year follow-up, plain x-rays showed no change in tumor size (Fig.2A,B). At two years, almost complete spontaneous regression of the tumor was observed (Fig. 3A,B), and the symptoms had disappeared.

Discussion

The most common benign bone tumor is osteochondroma[2,8]. The proximal humerus and the distal femur together account for almost 40% to 60% of all solitary lesions[2]. Its frequency of appearance is difficult to determine because of the large number of cases that remain asymptomatic. According to Mirra et al[9], small exostoses may be discovered fortuitously in 2% of patients in the course of a radiographic study for other reasons. This makes this lesion a very common entity, however, little is known about its natural history.

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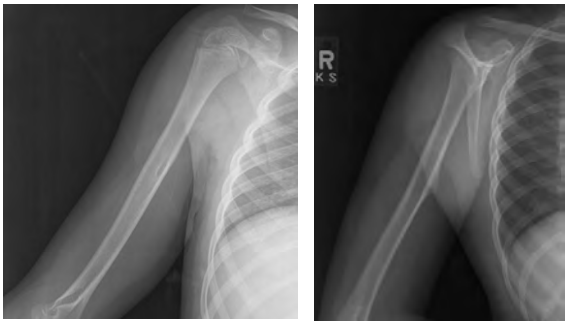
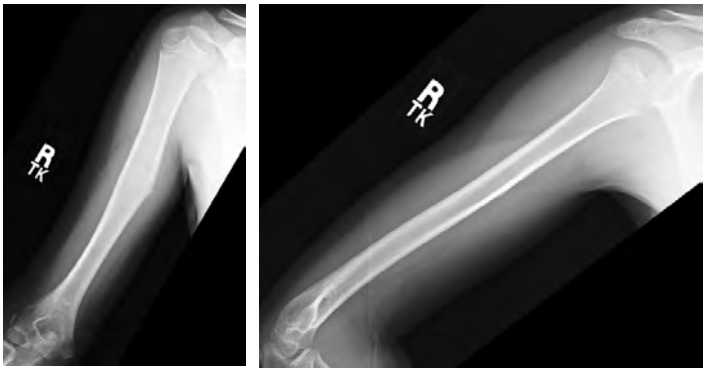
A B

Figure 1: Small bony exostosis arising from the anteromedial cortex, measuring approximately 2.7 cm at the base and projecting approximately 8 mm beyond the normal cortical line (A anteroposterior view; B, lateral view). This has a benign, well-corticated and typical appearance of an exostosis. The proximal and distal growth plates are normal. There is no evidence of focal bony destruction, soft tissue mass, or periosteal reaction.



A B

Figure 2. At 1 year from the diagnosis, comparable views (A anteroposterior view; B, lateral view). Osteochondroma of the mid-diaphysis of the humerus . There is no change in size in comparison with the previous ones. There is no evidence of focal bony destruction, soft tissue mass, or periosteal reaction.



A B

Figure 3. At 2 years from diagnosis, comparable views (A anteroposterior view; B, lateral view). Osteochondroma of the mid-diaphysis with interval development of cortical remodeling. The base does not appear as prominent as on the prior study and appears to be partially incorporated into the cortical thickening, measuring approximately 5 mm at the base and projecting 2 mm beyond the cortical normal line.

In 1835, Hunter first described spontaneous regression of an osteochondroma[10,11]. Since 1960, we identified 22 new cases of spontaneous regression of solitary osteochondromas (Table I)[8,11-28] reported in the literature.

Among the theories for spontaneous regression of a solitary osteochondroma are the one proposed by Copeland et al[20] and Castriota-Scanderberg et al[18] which says that the active resorption and metaphyseal remodeling caused by altered vascular supply due to fracture may lead to cessation of the cartilage cap and subsequent tumor regression. Paling et al[21] proposed that osteochondroma growth ceases before somatic growth ceases, followed by incorporation of the osteochondroma into the cortex by appositional growth of the adjacent bone. Choi et al[25] described resorption of osteochondroma as a result of an accompanying pseudoaneurysm.

Possible theories explaining the spontaneous regression include: the one documented by Copeland and Castriota-Scanderberg is related to the presence of a fracture and the one supported by Choi et al makes reference to the presence of a pseudoaneurysm.We do not think that these scenarios are related to our case . Paling's theory is related to a relationship among osteochondroma growth and somatic growth; in our case probably the osteochondroma growth ceased because our patient was near to her somatic growth cesation.Also, we can appreciate if we compare both sets of radiographs that there appears to be a partial incorporation of the osteochondroma into the cortical thickening.This is why we feel that Paling's theory explain better our case.

The most common documented locations involved in spontaneous regression of osteochondromas were the proximal humerus (7/22) and distal femur (6/22); this finding may be explained due to the higher incidence of osteochondromas in these locations.

The average age at the time of diagnosis was 8.8 years old (range, 5-15); 11.5 years old (range, 6 – 19) at the time of resolution and 2.8 years old (range, 0.16-6) until regression. Only two of the 22 patients had reached skeletal maturity at

Table 1. Spontaneous Regression of Solitary Osteochondromas – Review of literature.

Study	Year	Number of patients	Location	Type of osteochondroma	Age at diagnosis (y)	Time until regression (y)
Our case	2010	1	Middle third humerus	Sessile	11	2
Valdivielso-Ortiz et al[28]	2010	1	Distal femur	Sessile	9	4
Minami et al[11]	2009	1	Distal tibia	Sessile	6	2
Yasuda et al[27]	2008	1	Proximal tibia	Sessile	8	3
Hoshi et al[12]	2007	2*	Proximal humerus	Pedunculated	7	1.25
Arkader et al[8]	2007	1	Distal femur	Pedunculated	12	6
Choi et al[25]	2005	1	Distal femur	Sessile	12	0.16
Reston et al[13]	2004	1	Distal femur	Pedunculated	15	4
Yanagawa et al[14]	2001	1	Radius	Sessile	7	1.5
Yamamoto et al[15]	2001	1	Proximal phalanx hand	Sessile	3	6
Revilla et al[16]	1999	1	Proximal humerus	Sessile	9	5
Claikens et al[17]	1998	1	Distal ulna	Sessile	7	0.5
Castriota-Scanderberg et al[18]	1995	1	Proximal humerus	Sessile	5	2
Montgomery and LaMont et al[19]	1989	2	Proximal humerus/ proximal tibia	Sessile/pedunculated	10/11	5/3
Copeland et al[20]	1985	2	Distal femur	Pedunculated/sessile	11/10	2.5/2
Rosa et al[26]	1985	1	Distal femur	sessile	13	N/A
Paling MR[21]	1983	1	Proximal tibia	Pedunculated	9	1
Merle et al[22]	1980	1	Proximal humerus	Sessile	6	6
Callan et al[23]	1975	1	Proximal humerus	Sessile	5	1
Sellink JL[24]	1960	1	Proximal tibia	Sessile	8.5	3.5

y: years. * the second patient had Multiple Hereditary Exostosis (not included). N/A : No data available.

the time of regression. The oldest patient was fifteen years of age at the time of diagnosis and 19 years of age at the time of resolution.

Regression of an osteochondroma was recognized in 15 cases of the sessile type and 6 cases of the pedunculated type. As there is no good information as to the prevalence of sessile or pedunculated osteochondromas and as the number of cases of regression reported is small, no conclusion can be made as to the tendency toward regression by type of lesion.

Surgical resection of these bone tumors should be considered when the patient has persistent pain, cosmetic deformity, mechanical bone problems, continued growth, vascular or neurologic compromise and suspicion of malignant transformation[2,4,14,29-31]

Referred to as a simple surgical procedure, resection of an osteochondroma has been associated with some complications. Wirganowicz et al[30] performed a retrospective study in which a complication rate up to 12.5% was reported. Peroneal neurapraxia was the most commonly observed complication followed by arterial laceration, compartment syndrome, and fibular fracture. We have to take into account that this procedure can leave an important cortical defect that involves a risk of fracture, especially in the lower extremity, precautions must be taken, such as protective weight bearing with crutches for 6 weeks[5].

Another concern is malignant degeneration, which was first described in 1886; the risk is reported to be less than 1%, but may be much smaller[5,8]. Such malignant change is almost invariable toward chondrosarcoma, which develops on the layer of cartilage, although some rare cases of those that are of central location are the ones most frequently associated with this complication. If pain or continued growth occurs after reaching skeletal maturity, malignant degeneration should be ruled out by excisional biopsy[1,5,8,29].

The earliest changes occur near the cartilaginous cap and can be assessed by identifying an increase of the cap

thickness. Malignancy has been suggested when the cap is thicker than 2 cm in adults[3,29,32]. Magnetic resonance imaging (MRI), computed tomography and ultrasound have been used to measure the cartilage cap; many authors agree MRI is still the most accurate method to measure the thickness of the cartilage cap and to detect early malignant degeneration[3,32,33]. Most of these chondrosarcomas are grade I and is treated by means of broad surgical resection and limb conservation[5,34].

We report a patient with spontaneous regression of a solitary osteochondroma. It is possible this may occur with even higher frequency and, therefore, this possibility should be taken into account. A limitation in our case could be the x-ray projection; we have to bear in mind that if the x-ray beam is tangential to the osteochondroma, it could look smaller than it actually is.

This provides useful information for patients, families, and treating physicians and surgeons in discussing treatment options and deciding the most appropriate management. If solitary osteochondromas can regress by the age of 19 years, if less than 1% are going to become malignant and if none become malignant before the age of 20 years, then asymptomatic lesions could be observed for regression until fifteen to twenty years of age. This could safely reduce the need for and risks of unnecessary surgery for this tumor.

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