

ANGIOMA OF
THE MUSCLES OF THE CALF OF THE LEG AS
THE CAUSE OF PES EQUINUS

BY

HENRY JOSEFSSON

Primary angioma of striated muscle is not a particularly rare phenomenon, as Jenkins and Delaney in 1932 found 256 cases described in the literature, and these authors had even excluded about a score of doubtful cases. Since then, a few more cases have been described. Of these 256 cases, 59 showed some deformity or functional impairment of the extremity involved, and a tip-toe deformity was observed in 12 cases. As often it is difficult to establish clinically the cause of such pes equinus, and as a failure to notice the tumor may lead to symptomatic treatment instead of causal therapy, I wish to call attention to this genesis of pes equinus, in connection with the report of a case.

The case is that of a girl, 12 years old, who has been troubled with pain and tenderness of the calf of the left leg ever since the age of six years. The soreness was localized to the lateral side of the calf, and the child complained of it even on a fairly light touch—*e.g.*, washing of the leg, touching lightly the leg of a chair, etc. By and by, the parents noticed also some swelling of the calf.

This trouble persisted without any particular change, and after two years of illness (1930) her parents called the local doctor, who treated her with hot wet compresses. As 3 weeks of this treatment failed to give any improvement, she was sent to a hospital, where the diagnosis was made: Phlegmone anti-cruris sin. + lymphadenit. There was some swelling and redness of the left calf and oedema over the left tibia, besides some

tenderness on palpation of these parts. The inguinal lymph glands, on both sides, were pea-sized. X-ray examination showed no osseous changes in the upper half of the tibia and fibula. The temperature varied between 36.7 and 37.7. After one week of conservative treatment she was discharged as »feeling well«.

After her return home, the old trouble appeared anew. The tenderness over the calf persisted almost unchanged, but now also pain and tiredness of the calf would set in after exertion. On this account, for instance, she had to sit down and rest several times on her way to and from school. When sitting on a chair she nearly always pulled up her left leg (flexing in the knee and hip), as this position relieved the pain. The swelling of the calf varied, and the parents noticed it was less marked and more soft in the morning, larger and harder in the evening. Apart from this ache and tiredness, the child had no trouble in walking. In June 1934, the parents took her again to the local doctor who referred her to the Orthopedic Hospital of Härnösand under the diagnosis *pes equinus*.

Physical examination, July 16, 1934: Rather marked atrophy of the entire left leg especially below the knee. The left foot is fixed in maximal equinus position and cannot be flexed by passive motion. No varus position of the foot. The upper part of the calf is intensely tender to pressure. This tenderness is so marked that the girl has tears in her eyes and pulls up her leg abruptly even on very light palpation. The tenderness to pressure extends up to the region of the head of the fibula. In this region it is possible in the depth to make out an area of firmer consistency than the surroundings. No disturbances of the sensibility. In walking she puts only the toes of the left foot to the ground, on account of the equinus position. There is no pain when she stands on this foot. Moderate valgus position of the left knee. Full mobility of knee and hip joints. The legs are equally long. No other abnormalities revealed on examination.

X-ray examination of the leg shows no osseous changes, nor any definite changes in the soft part.

As the diagnosis was uncertain it was decided operatively to explore the palpable mass in the calf.

June 17: *Operation* (Dr. v. Rosen). Under ether anesthesia a longitudinal incision is made from the origin of the lateral head of the gastrocnemius down over the upper half of the calf. The lateral part of the muscles of the calf—in particular, as far as could be estimated, the soleus—was largely infiltrated by a spindle-shaped firm tumor, measuring about 20 cm. in length and 4 cm. in width. That part of the muscles which was infiltrated by the tumor was loosened from the deep musculature and extirpated. On examination of the extirpated mass, it looked questionable whether its upper part, *i.e.* at the origin of the muscle—contained any normal tissue. The extirpation did not give any bleeding worth mentioning. The extirpated tumor was bluish-red in color; on section, the cut surface showed many dark red spots. Plaster bandage, with the foot in the strongest dorsal flexion possible.

Two weeks later, open achillotenotomy was performed for further correction of the equinus position. Plaster bandage, with the foot in corrected position.

Microscopic exam. of tumor (Dr. Erik Laurén, Umeå): The specimen shows a cavernous hæmangioma invading the muscle in nearly its entire extent. The muscle fibres are markedly atrophic, embedded in connective tissue and, here and there, even in adipose tissue; there are some areas of round-cell infiltration. The upper part of the muscle shows no pronounced cavernous structure, but a rather abundant supply of thin-walled blood vessels; so the tumor may not have been removed completely. No sign of malignancy. On subsequent inquiry, Dr. W. Lundquist (Umeå), states that the walls of the cavernous tumor contained some elastic substance but, in his opinion, not any particularly great amount. Laborator N. Gellerstedt (Upsala), has been kind enough to point out for me the following interesting details in the histological picture. Between groups of cavernous blood spaces there are small accumulations of very muscular arterial branches, in part with entirely obliterated lumen, which may possibly constitute a component of the angioma itself (admixture of arterial racemose angioma). The other detail is the fact that several wide connective tissue septa

between the blood spaces contain streaks or small bundles of smooth muscle fibres without any visible connection with distinct blood vessels, and also islands or cords of muscle cells belonging to the striated system. Gellerstedt does not think this is a question of atrophic skeletal muscle fibres; he holds it for more likely that here we are meeting with embryonal muscle tissue.

The post-operative course was uneventful, and the patient was discharged on September 13, 1934, as being well.

Several reexaminations have been made subsequently, the last on September 4, 1935 when the following findings were recorded: The patient is now completely free from her previous trouble, and she has no difficulty whatever in taking part in all the work that is done on a farm; she also walks long distances without any inconvenience whatever. The examination reveals no faulty position of the foot; the mobility in the ankle is good. In full dorsal flexion there is still a deficit of 10 degrees. By passive motion the foot can be forced all way up. The power of plantar flexion is good. The contractions of the gastrocnemius are good, especially in the medial part of the muscle. The circumference of the calf is 4 cm. less and that of the thigh 1 cm. less than the corresponding measures of the right leg. She walks well, and she is able to stand on Tip-toe on the left foot alone, without any support. Sensibility normal.

In 1906 *Putti* described a case of primary muscular angioma of the calf of the leg as the cause of a pronounced deformity of the foot, and he considers this condition a new element in the neoplastic genesis of deformities. On several points his case is similar to the one described by me. Thus, the history of illness is very long. The pain in the calf and the limping began already in early childhood. Gradually there developed an extreme pes equinus with slight varus position. The patient, who is 33 years old had often been treated by various physicians. On palpation of the calf a peculiar resistance was noticed, apparently irregular in the depth of the calf. The leg was distinctly atrophic. Palpation of the calf and attempt at dorsal flexion were very painful to the patient. Roentgenography showed, among other

things, some depositing of lime salt in this area. Before the operation the diagnosis was neurofibroma. The tumor, which extended from the heads of the gastrocnemius to the tendon of the Achilles, was extirpated; in addition, achillotenotomy.

Putti describes at the same time another case of cavernous angioma that was not limited to the gastrocnemius and the muscle of the foot alone but involved also the gluteus maximus and medius. This patient showed not only a tip-toe deformity in the foot but also flexion of the knee besides abduction and rotation in the hip. The history of illness covered 20 years, and the pain was the predominant feature; in the later years there was also the increasing malposition of the extremity, which disabled the patient to such a degree that he was able to get about only by means of crutches. The correct diagnosis was made before the operation, which consisted in resection of one head of the gastrocnemius, together with achillotenotomy.

Rietkus has described a case of cavernous hæmangioma of the muscles of the calf of the leg and of the quadratus plantæ with equino-varus position of the foot and plantar flexion of the great toe. In this case the pain was very intense, and the malposition less pronounced. Partial excision of the tumor was performed, but a recurrence appeared before long; for this reason, the leg was amputated 8 months later.

Petersen, *Margarucci* and *Kolaczek* have each reported one case of angioma, in which the patients showed an extensor defect in the knee beside a tip-toe deformity. The angiomas were localized to the muscles of the calf of the leg; in *Kolaczek's* case the tumor also involved some muscles of the thigh.

Davis, *Davis* and *Kitlowski*, *Le Dentu*, *Murphy* and *Nagatomi* have each described one case of cavernous angioma localized to the gastrocnemius or to the soleus, or to both, with various degrees of pes equinus.

In a case reported by *Nast-Kolb* all the muscles of the foot and calf are involved by a diffuse cavernous angioma. The pre-operative diagnosis was angioliipoma. The leg was amputated, and yet there appeared a relapse in the amputation stump.

Several different theories have been advanced about the

genesis of primary muscular angiomas. Virchow holds that these tumors are due to a progressive dilatation of preexisting blood vessels with morbid processes in the vasa vasorum: The new-formation of the tumor then proceeds from these preexisting vessels through proliferation of vascular cords. Rokitanski and others think that angiomas develop separately from the vascular system, and that their connection with the vascular system is merely a secondary stage. Still, most authors, I think, look upon the hæmangioma as a congenital anomaly, growing through proliferation of preexisting blood vessels. Several authors think that traumatic injury plays a certain rôle in the further growth of an angioma.

Pathologic-anatomically the primary muscular angiomas may be divided into diffuse and circumscribed tumors. The diffuse are the more common. Having no definite borders, they infiltrate the surrounding tissue. They may even involve nerves, periosteum, bone, synovial membrane, large arteries and veins, besides the skin, even though this does not happen very often. The circumscribed muscular angiomas often show a sharp demarcation from the surrounding muscle, not infrequently they even have a fibrous capsule. As a rule, angiomas increase in size but very slowly. They are looked upon as benign tumors but as a rule they do not grow by benign expansion, but by infiltration. Some authors have even described metastases, but they are very rare phenomena. The same applies to a few cases in which there has been described a malignant degeneration.

The most frequent histological type is the cavernous angioma, consisting of irregular cavernous spaces lined with epithelium and filled with normal blood. The supporting framework consists of connective tissue, and the walls of the cavernous spaces often contain elastic fibres. The blood vessels entering into the tumor show changes in the walls. The striated muscle tissue may not show any change at all, or there may be changes ranging from simple atrophy to different degrees of degeneration. There may be bundles of smooth muscle fibres without any connection with blood vessels. Accumulations of round cells is a finding often mentioned. A detailed description of the histolo-

gical features of angiomas would fall outside the scope of this report.

Hæmangiomas may be found in any muscle, but they prefer the extremities, especially the lower. Of the 256 cases tabulated by Jenkins and Delaney, 84 were located in the trunk, 64 in the upper extremities, and 107 in the lower extremities; of the last-mentioned, 59 were found in the thigh, 39 in the calf of the leg. The larger and more essential muscles are involved more frequently. In general the tumor involves only one muscle (80 % of cases), but it may implicate two muscles or even a whole group of muscles. In a few cases such tumors have made their appearance in several muscle groups.

The sex distribution is equal. Angiomas occur as a rule in young persons. Thus 47 % of the cases appeared within the first 10 years of life, 79 % within the first 20 years, and 94 % within the first 30 years.

The symptoms are subject to great variation. Pain is the most common symptom; now and then it may be the first symptom, and it has been present in about 60 % of all cases. The pain may be localized to the site of the tumor, or radiate from the tumor out in the extremity involved. It may be elicited by pressure upon the tumor or by movement of the muscle involved. As a rule the pain subsides on resting. The pain may vary from a very dull ache to the most cutting attacks. There may be numbness of the extremity involved. Some authors hold that the pain is to be explained by the assumption that the tumor presses upon a nerve, or even that it infiltrates the nerve itself.

The tumor may vary in size from that of a hazel-nut to that of a child's head; it usually is palpable on thorough examination. Most often its consistency is somewhat elastic and spongy, but it may be very soft, and also hard. Sometimes the tumor is compressible, and this is a very significant sign. Changes in volume are not uncommon phenomena when large veins communicate with the cavernous spaces. The mobility of the tumor often varies with the flaccid or contracted state of the muscle. The overlying skin is usually free from morbid changes. If blood

is withdrawn in the syringe on puncture of the tumor, it naturally lends support to the diagnosis.

When hæmangiomas are located in muscles or muscle groups that are of particular functional importance, they give rise to conditions of functional insufficiency or to deformities. This holds true in particular if they are localized to the muscles of the extremities. Functional insufficiency and deformity are found in nearly one-fourth of Jenkins and Delaney's 256 cases. The functional disturbance may vary within wide limits. The contractile elements of the muscle are brought out of order by the angioma, causing thus a decrease in the functional capacity. Another cause of functional disturbance is the aforementioned pain that may make the involved extremity useless. Deformity is most frequent when the tumor is localized to the lower extremities. In those cases where pes equinus has been present the tumor is usually located in the muscles of the calf of the leg. As to the muscular insufficiency and deformity in cases of muscular angioma, Davis and Kitlowski say: »These tumors assume a malignant aspect when one considers that they inevitably terminate in the loss of function or sacrifice of an extremity if proper treatment is not carried out at an early stage«.

Generally the diagnosis is hard to make in the early stages, and in most of the cases described in the literature the diagnosis was made first in connection with the operation.

Patients with pes equinus due to muscular angioma seek medical advice either on account of the pain or because of the deformity. Careful and thorough palpation of the muscles of the calf may lead to the right diagnosis. In a few instances the diagnosis may be established by an explorative puncture. In most cases one has to be content to ascertain the presence of a resistance, differing in consistency from the surrounding muscles. There may be changes in the volume of the calf with changes in the position of the leg. The tumors grow but slowly. Most of the patients are under 20 years.

X-ray examination may be of some assistance, at least in cases where there are phleboliths or other forms of calcium

deposits in the tumor, and in the very rare cases where the periosteum is involved.

Because the muscular angiomas on palpation are often lobular or nodular, and because they grow so slowly, they are often mistaken for lipomas. Also sarcoma is a not uncommon erroneous diagnosis, but the angiomas generally grow more slowly. Hæmangioma may even be mistaken for fibroma, neuroma, neurofibroma, cold abscess, cysts, chronic myositis, gumma, »muscular hernis«, etc.

The prognosis is not always good, on account of irreparable deformities that may result when the diagnosis is made too late for a fully effective treatment. Relapse is reported in several cases.

The treatment consists in careful extirpation or excision of the tumor and, in the presence of pes equinus, achillotenotomy. Electrocoagulation, X-rays and radium treatments have been tried but without favorable result. Hæmorrhage during and after the operation is not uncommon. In a few instances it has been necessary to amputate the leg on account of relapse or wide extension of the tumor.

SUMMARY

Description is given of a case of pes equinus due to hæmangioma of the muscles of the calf of the leg in a girl of 12 years. The symptoms began at the age of 6 years with pain in the calf; later on the tip-toe deformity developed.

After a brief survey of previous cases described in the literature, an outline is given of the morbid anatomical picture of hæmangioma, with a brief mention of the variable localisation and symptomatology of these tumors. It is pointed out that in hæmangioma of the muscles of the calf it is difficult without explorative operation to ascertain the cause of the morbid condition. Not infrequently, however, an area of sore resistance is discovered in the calf on careful palpation. X-ray examination may at least in some cases corroborate the diagnosis. The patients are young. To avoid the risk of irreparable deformity

the treatment is to be instituted as early as possible, and it should consist in radical removal of the angioma.

ZUSAMMENFASSUNG

Verfasser beschreibt einen Fall von Spitzfuss bei einem 12-jährigen Mädchen, der durch ein Hämangiom in der Wadenmuskulatur verursacht war. Die Symptome begannen im Alter von 6 Jahren mit Schmerzen in der Wade, und später kam Spitzfussstellung hinzu.

Nach einem summarischen Überblick früher in der Literatur beschriebener Fälle von pes equinus mit ähnlicher Genese wird eine kurzgefasste Schilderung des pathologisch-anatomischen Bildes des Hämangioms gegeben, und dann werden die verschiedenen Lokalisationen und die wechselnde Symptomatologie behandelt. Betreffs der in der Wadenmuskulatur lokalisierten Hämangiome wird u. a. die Schwierigkeit hervorgehoben, die Krankheitsursache ohne operative Blosslegung zu konstatieren. Bei sorgfältiger Palpation der Wade findet man jedoch oft eine schmerzhaft Resistenz. Eine Röntgenuntersuchung kann, wenigstens in gewissen Fällen, die Diagnose sicherer gestalten. Die Patienten sind jung. Wegen des Risikos irreparabler Deformitäten soll die Behandlung, bestehend in radikaler Entfernung des Angioms, so früh wie möglich einsetzen.

RÉSUMÉ

Il est donné la description d'un cas de pes equinus dû à un hémangiome des muscles du mollet, chez une fillette de 12 ans. Les symptômes se manifestèrent à l'âge de 6 ans, par des douleurs dans le mollet; la déformation apparut plus tard.

Après avoir brièvement passé en revue les cas décrits antérieurement dans la littérature, il est donné un exposé du tableau anatomo-pathologique de l'hémangiome, avec mention sommaire des différentes localisations et de la symptomatologie de ces tumeurs. Lorsqu'il est question d'hémangiome localisé

dans la musculature du mollet, on relève entre autres qu'il est très difficile de déterminer les causes de la maladie, sans exploration opérative. En palpant minutieusement le mollet, on trouve cependant assez souvent une résistance douloureuse. Une radiographie peut, tout au moins dans certains cas, confirmer le diagnostic. Les malades sont jeunes. Afin d'éviter l'irréparable déformité, le traitement, qui consiste à enlever radicalement l'angiome, doit être pratiqué le plus tôt possible.

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