

Liposarkom – značaj kliničkog prepoznavanja od strane lekara opšte medicine

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Liposarcoma – the importance of clinical recognition by general physicians

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Sažetak

Uvod. Liposarkom je maligni tumor koji potiče iz masnih ćelija i drugi je najčešći sarkom mekog tkiva. Obično se javlja kod sredovečnih i starijih muškaraca (40 i više godina). Sarkomi spadaju u retke tumore. U Evropi se svake godine dijagnostikuje novih 4 do 5 slučajeva na 100.000 ljudi, bez značajne razlike među pojedinim zemljama. Pacijenti primećuju prisustvo bezbolne duboko usađene mase u mekom tkivu. Liposarkomiu najčešće lokalizovani na ekstremitetima, na retroperitoneumu, ređe u predelu glave i vrata. Većina bolesnika nema nikakve simptome sve dok tumor ne naraste toliko da počne da vrši pritisak na okolne organe, uzrokujući bol i poremećaj funkcije tih organa. Prognoza zavisi od tipa liposarkoma. Dijagnoza se postavlja na osnovu anamneze, kliničkog pregleda, CT-a (skenera), biopsije i patohistološke analize. Lečenje je hirurško uz adjuvantnu zračnu terapiju.

Cilj rada. Prikazom dva slučaja želeli smo da ukažemo na značaj lekara opšte medicine u ranom dijagnostikovanju liposarkoma, što doprinosi visokoprocenatnom izlečenju.

Prikazi slučaja. Pacijent muškog pola, 62 godine, promena na vratu, preko dve godine, postepeno raste, nakon operativnog tretmana, pH nalaz odgovara atipičnom, lipomatoznom tumoru/dobro diferentovani liposarkom. Pacijent ženskog pola, 77.godina, domaćica, više godina promena u kockicealnoj regiji koju je često operisala (sinus pilonidalis). Nakon operacije pH nalaz -epitheloid sarcoma/dobro diferentovan.

Zaključak. Svaki iskusni lekar opšte medicine, stoga, treba dobro uzetom anamnezom, detaljnim kliničkim pregledom, obraćanjem pažnje na alarmne simptome, uz razmišljanje i o diferencijalnoj dijagnozi i o ovoj vrsti ređih tumora pacijenta, da doprinese blagovremenom postavljanju dijagnoze i uspešnom rešavanju zdravstvenog problema.

Ključne reči: tumori mekih tkiva, liposarkom, primarna zdravstvena zaštita, porodična medicina, lekar opšte medicine.

Abstract

Introduction: Liposarcoma is a malignant tumor deriving from fat cells and the second most common sarcoma of soft tissues. It occurs most often in middle-aged and older men (over 40 years). Sarcomas are rare tumors. The incidence in Europe is 4-5/100.000 people per year and there is no difference between countries. A patient may notice a painless, deeply rooted mass in the soft tissue. They are mostly found on the extremities, retroperitoneum, and rarely on the neck and head. The majority of patients have no symptoms until the tumor reaches the size which may be pressuring the surrounding tissue, causing pain and organ dysfunction. Prognosis depends on the type of sarcoma. Diagnosis is based on medical history, clinical examination, CT scan, biopsy, and pathohistological analysis. The treatment is surgical with adjuvant radiation therapy.

Objective: We presented two case reports and the aim was to point out the importance of general physician (GP) in early diagnosing of liposarcoma, which may lead to a high percentage of healed patients.

Case report: A male patient, 62, presents with neck lump he has had for two years. It's been growing slowly. The patient was operated and the pathohistological findings showed it was atypical, lipomatous tumor/well-differentiated liposarcoma. A female patient, 77, housewife, suffers from a lump in the coccygeal area, which was operated on very often (considered to be sinus pilonidalis). After the last operation, a pathohistological analysis was performed and it was found to be epithelioid sarcoma/well differentiated.

Conclusion: Every GP should ask questions about the medical history, perform a clinical examination, pay attention to alarm symptoms, all the while bearing in mind differential diagnosis of this rare disease. Thus he may contribute to early diagnosing and successful treatment of the disease.

Keywords: soft tissue tumors, liposarcoma, primary health care, family medicine, general medicine.

Uvod

Liposarkom je maligni tumor koji potiče iz masnih ćelija i drugi je najčešći sarkom mekog tkiva. Često se javlja kod sredovečnih i starijih muškaraca (40 i više godina). Sarkomi generalno spadaju u retke tumore. U Evropi se svake godine dijagnostikuje novih 4 do 5 slučajeva na 100.000 ljudi, bez značajne razlike među pojedinim zemljama^{1,2}. Pacijenti obično primećuju prisustvo bezbolne duboko usadene mase u mekom tkivu. Ako se tumor povećava, može se pojaviti bol ili funkcionalno oštećenje. Liposarkomi su najčešće lokalizovani na ekstremitetima, na retroperitoneumu, ređe u predelu glave i vrata. Većina bolesnika nema nikakve simptome sve dok tumor ne naraste toliko da počne da vrši pritisak na okolne organe, uzrokujući bol i poremećaj funkcije tih organa. Liposarkom se uglavnom polako razvija, a bolesnik može katkad da primeti bolne otkoke, ukočenost, pojavu varikoziteta vena, bol u abdomenu, gubitak telesne težine, malaksalost, mučninu i povraćanje. Ponekad mogu postojati i neurološki simptomi i limfadenopatija. Prognoza zavisi od tipa liposarkoma. Dobro diferentovani i miksoidni tip imaju dobru prognozu, retko metastaziraju, dok nediferentovani i liposarkom okruglih ćelija recidiviraju i veoma brzo dolazi do metastaze. Dijagnoza se postavlja na osnovu anamneze, kliničkog pregleda, CT-a (skenera), biopsije i patohistološke analize. Lečenje je hirurško uz adjuvantnu zračnu terapiju³⁻⁷.

Cilj rada

Prikazom dva slučaja želimo da ukažemo na značaj lekara opšte medicine u ranom dijagnostikovanju liposarkoma, što doprinosi visokoprocenatnom izlečenju.

Prikaz slučaja

Slučaj 1. Pacijent MR, 62.godine, diplomirani ekonomista, direktor privatnog preduzeća, Dana 04.04.2018. godine javlja se radi promene na vratu koju ima preko dve godine, koja postepeno raste; ne navodi da je imao bolove niti crvenilo i svrab iznad kože, da samo ponekad ima osećaj zatezanja i da mu smeta. Želi da je operiše jer je primetio da od pre 12 meseci počinje rapidno da se širi. Lična anamneza: operacija žučne kesice 2003. godine., ima BHP (benigna hiperplazija prostate), oko pet godina, ali je pod kontrolom urologa, prima odgovarajuću simptomatsku terapiju, *Sy Gilbert*, nepušač, ne koristi alkohol, ne uzima nikakve lekove, nije hipertoničar. Porodična anamneza: majka i otac hipertoničari, negira dijabetes, astmu i maligne bolesti u porodici. Opšte stanje - uobičajene osteomuskularne građe. TT 90 kg, TV 185 cm, afebrilan, TA 120/80 mmHg, EKG: sinusni ritam, srčana frekvencija 70/min, levogram, redukovani R u D₃ pri auskultaciji nema šuma nad srčanim ušćima, pulmo: pri auskultaciji normalan, disajni šum bez propratnih patoloških

Introduction

Liposarcoma is a malignant tumor deriving from fat cells and the second most common sarcoma of soft tissues. It occurs most often in middle-aged and older men (over 40 years). Sarcomas are rare tumors. The incidence in Europe is 4-5/100.000 people per year and there is no difference between countries^{1,2}. A patient may notice a painless, deeply rooted mass in the soft tissue. They are mostly found on the extremities, retroperitoneum, and rarely on the neck and head. The majority of patients have no symptoms until the tumor reaches the size which may be pressing on the surrounding tissue, causing pain and organ dysfunction. Liposarcoma develops rather slowly and a patient may notice painful swelling, stiffness, varicose veins, abdominal pain, weight loss, fatigue, nausea, and vomiting. Sometimes, neurological symptoms and lymphadenopathy may appear. Prognosis depends on the type of liposarcoma. The well-differentiated and myxoid type have got a good prognosis, rarely metastasize, while undifferentiated and round cell liposarcoma relapse and metastasize very fast. Diagnosis is based on medical history, clinical examination, CT scan, biopsy and pathohistological analysis. The treatment is surgical followed by adjuvant radiation therapy³⁻⁷.

Objective

We presented two case reports and the aim was to point out the importance of general physician (GP) in early diagnosing of liposarcoma, which may lead to a high percentage of healed patients.

Case report

Case 1: A male patient, 62, an economist, CEO in a private firm, presents on April, 4th, 2018 with a neck lump he has had for two years and it has been growing slowly. He has no pain, redness, itchiness of the skin. Occasionally he feels tightness in the area and it bothers him. He wants it removed because he noticed the lump growing rapidly in the last year. Personal medical history: cholecystectomy in 2003; suffers from BPH for five years now and is seeing a urologist regularly; also uses symptomatic therapy; *Sy Gilbert*, doesn't smoke or use alcohol, doesn't suffer from hypertension, doesn't take any other medications. Family medical history: Mother and father suffer from hypertension. Physical findings: of average osteomuscular structure, height 185 cm, weight 90 kg, afebrile, BP 120/80 mmHg, ECG: sinus rhythm, puls 70 beats per minute, left axis deviation, reduced R wave in D₃, lungs clear to auscultation; normal heart rhythm, no heart murmurs or thrills; abdomen soft and non-tender. There is a lump on the back of his neck (regio nuchae).

šumova, abdomen pri palpaciji uredan nalaz, ostali nalaz po sistemima uredan. Kliničkim pregledom uočava se promena na vratu u predelu potiljačne (*nuhalne*) regije, ali se u odnosu na medijalnu liniju proteže ulevo i udesno. Promena je 12 cm -15 cm ali neravnomerno ograničena, izgleda razliveno, koža neizmenjena, pri palpaciji nema bola, meke konzistencije na nekim delovima, stiće se utisak fiksiranosti za podlogu. Palpacijom regionalnih limfatika nema uvećanja niti bola. Pacijent upućen na osnovne laboratorijske analize, Ultrazvuk (UZ) regije vrata gde se nalazi promena i UZ gornjeg abdomena. Laboratorijski. nalazi uredni, urin uredan, šećer u krvi (šuk) 6,0 mmol/l, kreatinin 68 mmol/l, bilirubin 39,5 mmol/l, holesterol 5,6 mmol/l, UZ abdomena uredan nalaz; UZ vrata u predelu potiljačne regije (*regio nuchae*) supkutano od medijalne linije prema lateralno, uočava se hipo do izoechogena promena, jasno ograničena, delimično inkapsulirana promera oko 19 mm koja po svojim karakteristikama odgovara lipomu; u potiljačnoj regiji od medijalne linije prema lateralno levo identična promena septirana, prevazilazi mogućnost merenja sonde, pa nema tačnog promera, ali lipomu odgovara i submandibularno desno identična promena oko 8 mm koja, takođe, odgovara lipomu. Pacijent je upućen na pregled kod plastičnog hirurga radi dalje dijagnostike i eventualnog hirurškog odstranjenja (ekstirpacije) promene pod dijagnozom *Lipoma in obs* MKB10 D17. Dana 24.08.2018. godine pregledan od strane plastičnog hirurga i zbog opsežnosti promene zakazana hospitalizacija za 20.09.2018. i savetovana preoperativna priprema pacijenta; 26.09.2018. godine pacijent dolazi sa otpusnim listom, lečen na KCV Novi Sad – Klinika za plastičnu i rekonstruktivnu hirurgiju 20-24.09.2018. godine i urađena *extirpatio in toto* - promene (veće promene promera 19 mm) i poslat nalaz na pH analizu; 03.10.2018. dobija pH nalaz: 1589/18 - MKB10 C49.0, koji odgovara atipičnom, lipomatoznom tumoru/dobro diferencijovanom liposarkomu (*atypical lipomatous tumor/well differentiated liposarcoma*) - *lipoma like* (nalik lipomu). Radi daljeg lečenja, pacijent upućen na preoperativni zahvat - *extirpatio radicalis*. Lečen 11-18.10.2018. zatim upućen na pregled Onkološkoj komisiji za meka tkiva Instituta za onkologiju IOV Sremska Kamenica - radi dalje terapije; 27.12.2018. nalaz Onkološke komisije: dalje indikovani režim redovnih kontrola kod nadležnog onkologa, hirurga i dermatologa, kontrola Onkološke komisije za tri meseca sa nalazima laboratorije: UZ vrata i regije od interesa, UZ abdomena i RTG pluća, savet za strogu zabranu izlaganja sunčevim zracima i ECOG score 0. Kontrola plastičnog hirurga 16.01.2019. – nema znakova lokalnog recidiva, subjektivno pacijent bez tegoba i dalja kontrola predviđena za tri meseca, UZ nalaz uredan; nalaz na mestu ekstirpacije tumora: uočava se anechogena promena tipa seroma, ostali nalaz uredan. Pacijent je dobrog opšteg stanja, nije gubio na težini, kliničkim pregledom nema patoloških promena niti znakova recidiva i savetuju se dalje kontrole.

The size of the lump is 12-15 cm, it has irregular margins, no changes on the skin, not tender to palpitation, soft consistency in some parts, fixed to the base. Regional lymph nodes are not enlarged or painful. The patient did lab results and ultrasonography of the neck and abdomen. Lab results were insignificant (urine-normal; glucose-6.0 mmol/l; creatinine-68 mmol/l; bilirubin-39.5 mmol/l; cholesterol 5.6 mmol/l). Ultrasonography of the abdomen was normal but on the back of the neck (*regio nuchae*) there was a subcutaneous tumor spreading laterally, on both sides. It was hypo to isoechogenic, with clear margins, partly encapsulated, 19 cm in diameter and its characteristics were consistent with lipoma. In the nuchae region, on the left lateral side, there was an identical septated formation, which couldn't be measured (exceeds the measuring ability of the probe), but is also consistent with lipoma. In the submandibular region, there was also a formation, 8 mm in diameter, lipoma-like. The patient was referred to a plastic surgeon for further evaluation (Diagnosis – *Lipoma in obs*. MKB10 – D17). A plastic surgeon examined him on August, 24th, 2018 and due to the extension of the tumor he scheduled the hospitalization for September, 20th, 2018. On September, 26th, 2018 the patient visited his GP and brought a discharge letter. He was hospitalized at the Clinic for plastic and reconstructive surgery, of the Clinical Center of Vojvodina, Novi Sad from 20.- 24.09.2018. where the total removal of the bigger tumor was performed and it was sent to pathohistological analysis. The PH analysis arrived on October, 3rd, 2018 – Diagnosis: C49.0 - *Atypical lipomatous tumor/well-differentiated liposarcoma* - *lipoma like*. His further treatment included new operation (*extirpatio radicalis*) which was performed at the same clinic and he stayed at the hospital from 11.-18.10.2018. After surgical treatment, the patient was referred to the Oncology commission for soft tissues of the Oncology Institute of Vojvodina, Sremska Kamenica for further treatment instructions. The commission instructed further follow up by an oncologist, surgeon, and dermatologist. The patient was supposed to check in with the commission in three months with new lab results, ultrasonography of the neck and abdomen, and lung X-rays. He was advised to forego sun exposure and his ECOG score was zero. On his follow up with a plastic surgeon, on January 16th, 2019 he was told the disease wasn't relapsing and personally he felt good. So his next follow up was scheduled in three months. Ultrasonography of the neck showed anechogenic formation, consistent with seroma, in the operation area. Considering his overall good health and no signs of relapse he was advised just to follow up on his scheduled check-ups.

Slučaj 2. Pacijentkinja MB. 77.godina, domaćica, javlja se 21.09.2018.godine radi bolova u sedalnom delu leđa; navodi da ima promenu u vidu zagnojenog masnog tkiva poslednjih 6 meseci, ali da joj nije smetalo. Pacijentkinja takođe navodi da od pre nekoliko godina ima promenu na istom mestu, koju je često operisala, navodi da je u pitanju bio upaljeni *sinus pilonidalis*, ali nema nalaze ranijih operacija. Sada do-lazi sa osećajem slabosti i malaksalosti, izraženim bolom u sakralnom delu kičme, oseća bol na dodir i bol kada se kreće i kada sedi. Objektivnim pregledom, u predelu iznad glutealne brazde u sakralnom delu kičme uočava se palpabilna masa veličine 10 cm x 10 cm, koja je bolna na dodir, nejasno ograničena od okoline, ne može se pregledom oceniti njena dubina, površina iznad promene je crvena i ima znakovna fluktuacije. Lična anamneza: objektivno gojazna, TT 80 kg, TV 165 cm, TA 120/80 mmHg, EKG b.o., pulmo: auskultatorno normalan disajni šum, povremeno čujan niskotonski vizing, polimorfni niskotonski zvižduci koji se gube nakon kašlja, pušač, astmatičar oko 10 godina, ima redovne kontrole pulmologa i terapiju (*Seretide disc i Berodual pp*); laboratorijski nalazi: KKS b.o., šuk 6,2 mmol/l, rađena i kontrola OGTT- uredan nalaz, ginekološki b.o., Porodična anamneza: majka hipertoničar, lečena od karcinoma dojke. Pacijentkinja se zbog izraženih znakova akutne upale u pravcu *susp. Sinus pylonidalis* šalje 22.09.2018. godine na pregled kod opšteg hirurga radi saniranja promene. Urađena *extirpatio in toto* i materijal poslat na pH analizu. Rana načinjena nakon operacije se previja svaki dan uz primenu antibiotske terapije i uredno zarasta. Pacijentkinja 29.10.2018. dolazi na kontrolu sa nalazima pH: *Epitheloid sarcoma* - dobro diferentovan, jasno ograničen, MKB DG C44.7. Pacijentkinja je 06.11.2018. godine upućena na Onkolosku komisiju - internisti onkologu. Radi dalje dijagnostike i terapije, 8.11.2018.donosi nalaz Onkoloske komisije: savetuje se radi dalje kontrole praćenje stanja pacijentkinje na tri meseca: UZ regije od interesa i MR karlice, jer je opšte stanje pacijentkinje uredno; a UZ nalaz na kontroli ukazuje na ekstirpaciju promene u celosti bez znakovna širenja tumora; UZ abdomena i male karlice je uredan, lab nalazi uredni. Pacijetkinja 28.01.2019. dolazi sa nalazima onkologa interniste - rađen je kontrolni UZ regije od interesa, UZ abdomena, RTG pluća i MR karlice i nalazi su uredni, nema znakovna recidiva tumora niti znakovna širenja procesa; laboratorijski nalaz bo, šuk 6,2 mmol/l. Pošto je i gojazna, savetovano je da se uputi endokrinologu radi dalje dijagnostike u pravcu insulinske rezistencije. Kontrola onkologa sa istim nalazima se ponavlja i 20.03.2019. i potom 24.06.2019. i nalazi su uredni. Pacijentkinja je dobrog opšteg stanja, kontrole onkologa se savetuju na 6 meseci, kao i regulisanje telesne težine i dalji pregledi endokrinologa, jer je dijagnostikovana i insulinska rezistencija, dat savet za kontrolne mamografije jednom godišnje zbog pozitivne porodične anamneze.

Case 2: A female patient, 77, a housewife, presents with the pain in the gluteus area due to the festered fat tissue formation. She had it for 6 months but it didn't bother her until now. She actually had this lump for several years and it was often operated on (considered to be inflamed sinus pilonidalis) but has no medical records. Now she feels weak and exhausted and has severe pain in the sacral area. The pain appears on palpation and when she moves and sits. On examination, a palpable mass 10x10cm was found in the gluteal cleft. It's tender to palpation, with irregular margins. Its depth couldn't be determined but the skin above it was red and it showed the signs of fluctuation. Personal medical history: obese, weight 80kg, height 165 cm, BP 120/80 mmHg; ECG findings were normal, lungs were clear to auscultation, with occasional low-pitched wheezing which disappeared after coughing. She is a smoker and has had asthma for ten years. She has regular check-ups with her pulmonologist and uses asthma inhalers (*Seretide disc and Berodual inhaler*). Her lab results were in the normal range (CBC-normal count, glucose-6.2 mmol/l, glucose tolerance test was normal). The gynecological exam was also normal. Family medical history: her mother has hypertension and breast cancer. Inflamed sinus pilonidalis was suspected and she was referred to a surgeon. The surgeon performed an *Extirpatio in toto* on September 22nd, 2018 and sent the material for pathohistological analysis. The wound was dressed daily and she was prescribed an antibiotic. On October 29th, 2018 she presents with her pathohistological findings – *Epitheloid sarcoma*-well differentiated, with clear margins (MKB10 – C44.7). On November 6th, 2018 she was referred to the Oncology commission for further evaluation. She was advised to come for a follow up in three months with ultrasonographic findings of the operated area and pelvic MRI. This was due to her otherwise good overall health and her ultrasonographic and lab results showed no signs of relapse. On January 28th, 2019 she had a scheduled check-up with her oncologist and her findings were normal with no signs of relapse (ultrasonographic exam of the operated area and abdomen, lung X-rays, pelvic MRI). Since she is obese and has elevated blood sugar she was referred to an endocrinologist to evaluate her insulin resistance. She visited her oncologist on March 20th, 2019 and June 24th, 2019 and both times her findings were normal. She feels well and oncologist follow-ups were advised every 6 months. She was also advised on a healthy diet and weight reduction and to follow up on her endocrinologist check-ups since she was diagnosed with insulin resistance. She was also advised to schedule a mammography screening due to her family medical history.

Diskusija

Upravo zbog mnogo veće učestalosti benignih lezija mekih tkiva, koje su daleko više i češće rasprostranjene od sarkoma jer je diferencijacija između njih teška, pogotovo kliničkim pregledom - postavlja se pitanje koje pacijente treba uputiti na dalje dijagnostičke preglede. Veoma je važna dobro uzeta anamneza, koja se odnosi na porodičnu anamnezu u smislu češće pojave malignih bolesti u familiji - retinoblastom, neurofibromatoza tip 1 (*Recklinghausen*), tuberozna skleroza, familijarna adenomatozna polipoza (FAP), Li-Fraumeni sindrom, Werner sindrom, sindrom nevusa bazalnih stanica, ranije bolesti, faktori rizika za nastanak maligniteta, povrede, izlaganje sumnjivim i potencijalno kancerogenim materijama. Potom, značajan je detaljan klinički pregled promene koja se odnosi na češće lokalizacije tipične za sarkome, i promene koje su veće od 5 cm u prečniku ako pri palpaciji leže dublje u tkivima, nemaju jasno definisane ivice (tipa razlivanja), fiksirane za podlogu i alarmne simptome pacijenta (mršavljenje, malaksalost, promena kože). Prema ESMO smernicama kliničke prakse, sarkomi mogu duže vreme postojati bez bilo kakvih simptoma bolesti, a kada se oni i jave zavise od dela tela koji je zahvaćen^{1,6,5}. Značajno je da dugo *miruju* i da potom u kratkom periodu (1-2 god.) kreću nagli rast tumorozne promene. Bitna je stoga i odgovarajuća rana dijagnostika tipa radiološkog pregleda, kako bi se procenila proširenost bolesti i utvrdilo prisustvo ili odsustvo udaljenih metastaza⁴. Radi detaljne dijagnoze potrebno je uraditi biopsiju tumora za laboratorijsku analizu i potvrdu dijagnoze, kao i da bi se dobilo više informacija o tipu sarkoma jer od toga dalje zavisi prognoza i terapija bolesti^{2,3,7}.

Zaključak

Svaki iskusni lekar opšte medicine dobro uzetom anamnezom, detaljnim kliničkim pregledom, obraćanjem pažnje na alarmne simptome, uz razmišljanje o diferencijalnoj dijagnozi i o ovoj vrsti ređih tumora pacijenta, treba da doprinese blagovremenom postavljanju dijagnoze i uspešnom rešavanju zdravstvenog problema.

Discussion

Due to a much higher incidence of benign tumors of soft tissues, it is very difficult to differentiate which one is a sarcoma, especially during the physical examination. It remains to be seen which patients should be referred to further diagnostic procedures. Patient's family medical history concerning malignant diseases is very important (family history of retinoblastoma, neurofibromatosis type 1 – *Recklinghausen*, tuberous sclerosis, familial adenomatous polyposis, Li Fraumeni syndrome, Werner syndrome, basal cell nevus syndrome), earlier disease, risk factors for malignant diseases, previous injuries, exposure to potentially cancerogenic substances. Medical examination of the lump is also of great importance, especially if it's located in predilection locations for sarcomas, if it's larger than 5 cm in diameter, if it's deeply rooted in the tissue, has irregular margins (spilling effect), fixed base, with alarm symptoms (weight loss, fatigue, skin changes). According to ESMO guidelines, sarcomas could be asymptomatic for quite a while and the appearance of the symptoms often depends on the tumor location^{1,6,5}. They may be silent for a very long time and then in a short period of time (1-2 years) start growing rapidly. Early diagnosis is important and it should include a radiology exam to evaluate the disease spreading and existence or absence of metastases.⁴ For the final diagnosis it is important to perform a biopsy of the tumor which could give us more information on the type of the sarcoma. Prognosis and therapy depend on the biopsy findings.^{2,3,7}

Conclusion

Every experienced GP should be able to make a timely diagnosis and contribute to successful problem solving by acquiring a patient's medical data, performing a thorough physical exam, paying attention to alarm symptoms, all the while bearing in mind differential diagnosis and these rare tumors.

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