ANNALES
UNIVERSITATIS MARIAE CURIE-SKŁODOWSKA
LUBLIN – POLONIA
VOL. LXII, N 1, 65 SECTIO D 2007

1st Department of Radiology, Medical University of Lublin

PIOTR ORZECHOWSKI, ELŻBIETA CZEKAJSKA-CHEHAB, ANDRZEJ DROP

Multiple angiomyolipoma of the liver and kidneys diagnosed with multislice computed tomography – a case report

Angiomyolipoma (AML) belongs to rare, unencapsulated, benign tumours of the liver with higher incidence in females. It consists of three basic elements: smooth muscle cells, thick-walled blood vessels and fatty tissue, whose proportions vary from 5–90%. Histopathologically, AMLs may be divided into lipomatous (> 70% of fat), myomatous (<10% of fat) and pseudotumour-like types. The pseudotumour-like types are subdivided into angiomatous, pelioid or inflammatory (17). AML occurs in two clinical forms – as an isolated variation or a tumour coexisting with tuberous sclerosis complex (TSC, Bourneville's disease). Clinical examinations show that single or multiple angiomyolipomas of the liver are found in 6–24% of patients with tuberous sclerosis complex (4, 12, 13).

In the Bourneville's disease, the changes in the brain and internal organs are accompanied by dermatological manifestations – *adenoma sebaceum*. The disease is inherited as an autosomal, dominant feature. The characteristic grey-white nodules are observed in the cerebral cortex and ventricular regions. Various lesions develop in the internal organs: in kidneys – polycystic renal degeneration and angiomyolipomas in about 80% of cases, in lungs – cystic lesions, in the heart – myomas, in the liver – lipomas or angiomyolipomas, in bones – cysts and cartilaginous exostosis, in ovaries – cystic lesions (5, 6, 9).

The authors present three cases of AML of the liver and coexisting pathologic lesions diagnosed using multislice computed tomography (MSCT).

All the examinations were carried out with collimation 1.2 or 2.5 and 5 mm 8-row CT scanner GE LightSpeed Ultra using native scans and the multi-phase method in the arterial, parenchymatous and delayed phases (2 cases) after intravenous administration of the contrast medium. The contrast medium (100 ml Ultravist 300, Schering, Germany) was administered with the power injector at the speed of 3 or 4 ml/s. The arterial phase delay was determined by the bolus tracking technique. In all the cases the area examined involved the abdomen and supradiaphragmatic regions, in one case – the pelvis was also included.

CASE 1

A 38-year-old female patient (A.L.) hospitalized in the Department of Gastroenterology was sent for CT due to US-detected focal lesions and heaviness-like complaints. The US examination revealed several well-delineated, hyperechogenic foci typical of haemangiomas, 7–39 mm in diameter (Fig. 1). Fifteen years earlier the US scan showed the uneven contour and enlargement of the lower pole

of the right kidney with the blurred corticomedullare structure interpreted as the tumourous lesions and two hyperechogenic foci in the liver defined as haemangiomas. In the same year the patient underwent right nephrectomy, the postoperative diagnosis was the atypical, benign mesenchymal tumour, most probably angiomyolipoma.

MSCT showed two bigger foci in the liver – 32.8 mm and 27 mm. as well as eight smaller ones – 1–8 mm, density from (-)32 to (-)80 HU (Fig. 2). In the retroperitoneal space the presence of the area located medially to the inferior main vein and at the level of the left renal vessels of similar density was found whose size was 9x30x11 mm. Another lesion, 20x48x12 mm, density (-)108 HU, was detected in the soft tissues of the back on the right side (Fig. 3). The mean density of the foci described following the contrast medium was about (-)60 HU; in big foci and the lesion observed in the soft tissues the linear, irregular structures of higher density were visualized. The MSCT scan of the liver nodules was characteristic of angiomyolipoma, while the lesion in the retroperitoneal space and integuments was interpreted as lipoma. In the supradiaphragmatic region of the lungs single, thin-walled cysts were found, up to 7 mm in diameter, surrounded by normal parenchyma (Fig. 4). No lesions were detected in the site of kidney removal.

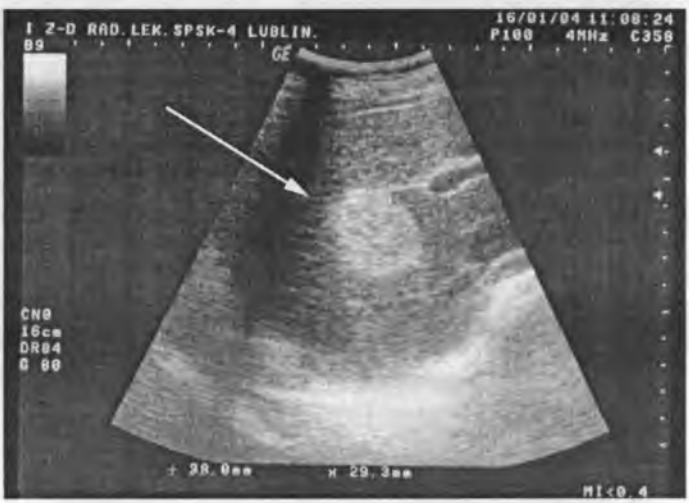

Fig. 1. A.L. In US examination, a hyperechogenic lesion in the liver is visible

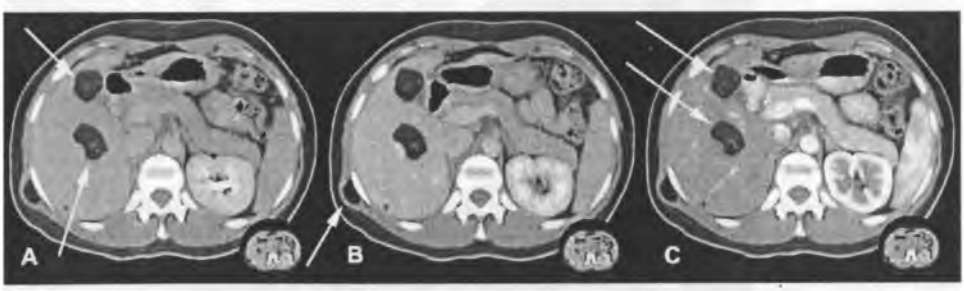

Fig. 2. A.L. MSCT axial view shows focal lesions of angiomyolipoma of low density with slight central enhancement in the arterial (A), portal (B) and delayed (C) phases.
A typical lipoma is visible in the soft tissues of the back on the right side

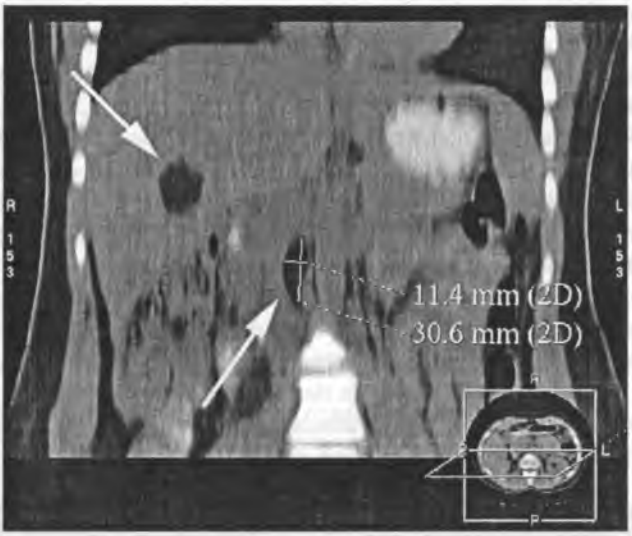

Fig. 3. A.L. MSCT – coronal reconstruction shows angiomyolipoma of the liver and fatty tissue area on the right side in the retroperitoneal space interpreted as lipoma

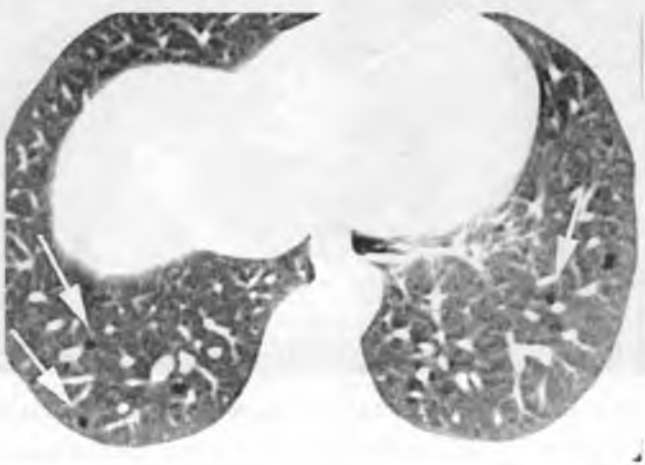

Fig. 4. A.L. MSCT – axial view in pulmonary window shows multiple small cystic lesions typical of LAM

CASE 2

A 23-year-old female patient (M.G.) of the Department of Urology was sent for CT because of unclear picture of renal changes which were earlier assessed using axial CT. MSCT showed abnormal contours of both kidneys with nodular irregular structures of heterogenic density. The biggest tumour – 50x45x55 mm, was located in the right kidney, its density was mostly typical of the fatty tissue (Fig. 5). In the left kidney the analogical lesion – 30x35x32 mm, was detected. The foci were strongly enhanced in the arterial phase; their densities were high in the parenchymal and delayed phases (above 80 HU). Moreover, several round foci up to 15 mm in size and of 2–20 HU

density were visible in both kidneys, which were mainly located subcapsularly. These changes in the subsequent phases of the examination became partially peripherally enhanced, which might have suggested that their origin is similar to that of the lesions in the upper poles. The density of some foci was typical of cysts; however the US scan showed their hyperechogenicity, which suggested angiomyolipoma. Furthermore, the liver was found to be moderately enlarged with eight foci, up to 5 mm in diameter and density from (-)30 to (-) 80HU, whose US appearance was typical of haemangiomas. The remaining abdominal organs were normal; however two pathologic changes of density and enhancement typical of lipoma were detected in the epicardium near the apex of the heart (28x19x36 mm) and in the lower part of the interventricular septum (18x10x7 mm) (Fig. 6). During the assessment multiple osteosclerotic foci – 4–9 mm, were found in the bony window located in the upper body of the spinal vertebrae (Fig. 7).

The patient was diagnosed with multiple hamartomatic lesions and qualified for close clinical observation. Two follow-up CT examinations performed at yearly intervals did not reveal any progression of the lesions.

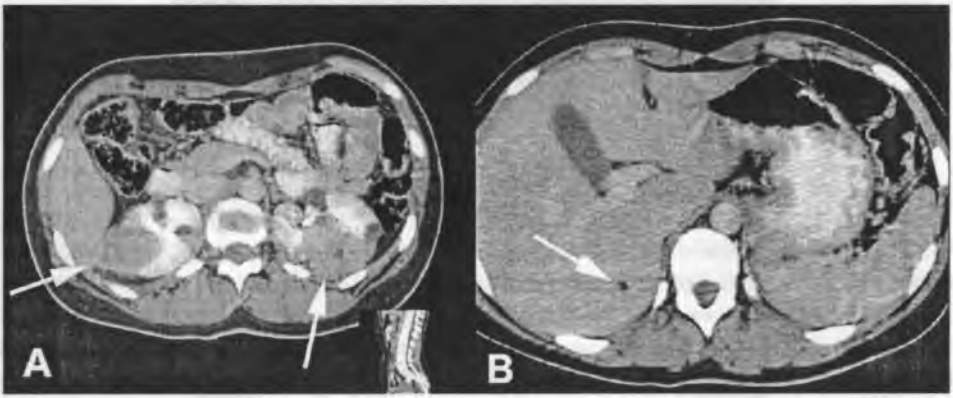

Fig. 5. M.G. MSCT in the excretory phase shows well-vascularized, multiple tumors of kidneys, contained fatty tissue and one of the small liver nodules of fatty density

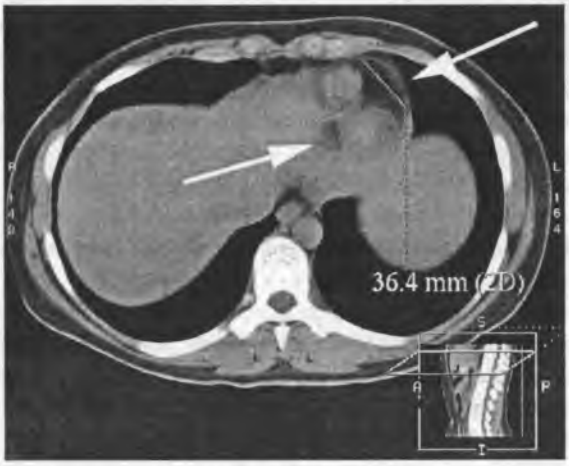

Fig. 6. M.G. MSCT reveals lesions of lipomatosis in the region of the heart apex and in the lower part of the interventricular septum

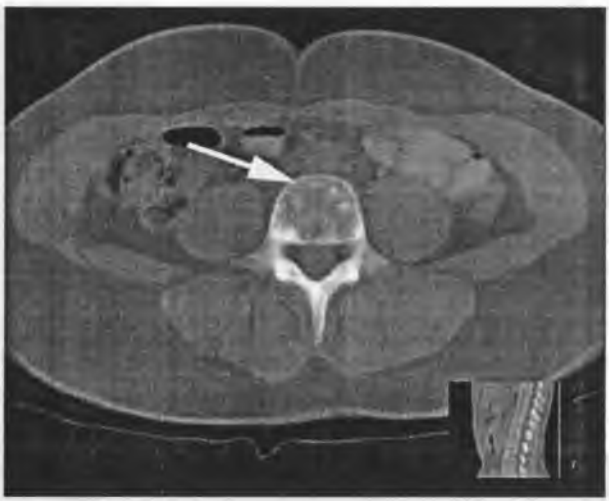

Fig. 7. M.G. MSCT in bone window shows multiple sclerotic lesions of the body of lumbar vertebra

CASE 3

A 23-year-old female patient (A.A.) hospitalized in the Department of Urology was sent for CT with the diagnosis of extensive nodular changes in the kidneys. MRI of the head carried out earlier demonstrated periventricular nodules typical of tuberous sclerosis.

MSCT revealed enlarged and nodularly changed kidneys. The intestinal loops with mesenteric vessels were shifted to the left and down towards the left iliac fossa. The extensive nodular lesion of the right kidney was 260x180x120 mm in size and extended downwards to the pelvis; on the left side it bordered the left kidney.

The left kidney contained the nodular lesion, 90x70 mm in its upper pole and smaller tumours, 35 mm in diameter in the remaining parenchyma (Fig. 8). In the right kidney the nodular lesions involved almost all parenchyma, while in the left kidney the disseminated tumour was found in the upper pole and well-delineated tumours of various sizes were detected in the lower and medial part with contrast enhancement of the remaining parenchyma in the individual phases of the examination. The right kidney tumour contained numerous, irregular fatty tissue areas coexisting with solid cystic lesions. The structure of tumours showed solid regions well supplied with blood with widened, winding vascular malformations. The main arterial and venous branches of the kidney vessels were shifted, pyelocalyceal systems widened and deformed by the compressing pathological tissue. A small amount of fluid was detected at the lower pole of the right kidney, additionally trace amounts of fluid were observed in the peritoneal cavity.

Four foci of negative density 7–27 mm in diameter, with linear internal structures, enhanced in the postcontrast phases were demonstrated in the hepatic parenchyma. In our opinion, CT scan showed extensive hamartomatic angiomyolioma-like lesions in the liver and both kidneys.

In the supradiaphragmatic region the following changes were visualized: moderately enlarged heart, features of left-side pneumothorax, numerous, fine cysts and parenchymal ground glass opacity (Fig. 9). The supradiaphragmatic segments on the left side showed also a relatively extensive area of pulmonary tissue consolidation which might correspond to inflammatory-atelectatic changes. Additionally, well-separated osteosclerotic foci of various sizes were observed in the bodies and arches of spinal vertebrae, as well as in the sacral bone.

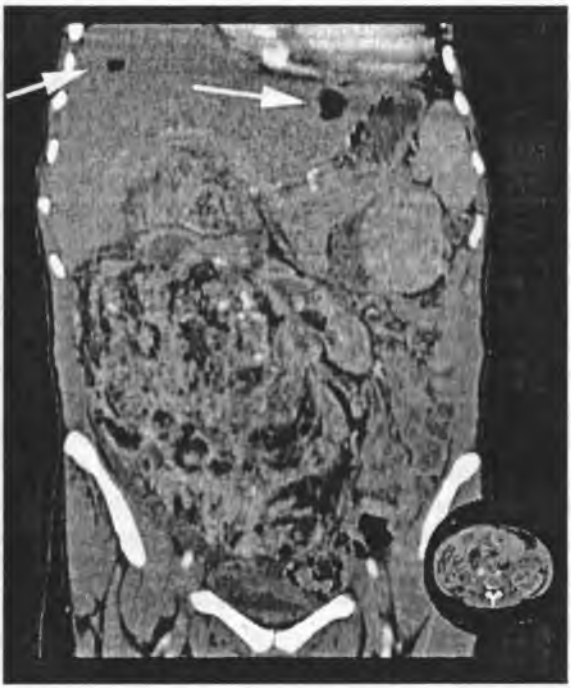

Fig. 8. A.A. Patient with tuberous sclerosis complex. The frontal reconstruction reveals giant, bilateral angiomyolipomas of the kidneys, two lesions of angiomyolipoma in the liver and a small amount of fluid in the pelvis

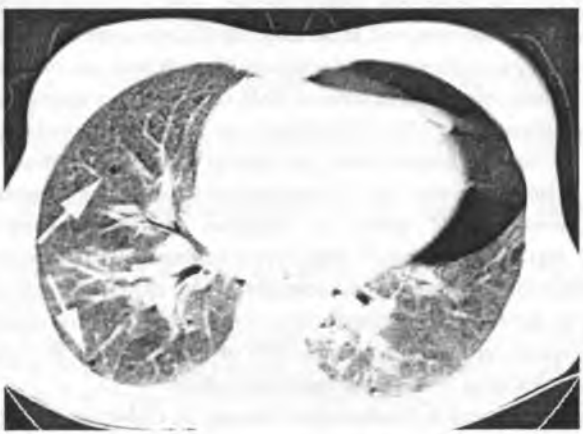

Fig. 9. A.A. MSCT in pulmonary window shows small cysts in the pulmonary parenchyma and the left pneumothorax detected by CT

DISCUSSION

The detection and differential diagnosis of AML of the liver are usually based on ultrasound and CT examinations; MRI is also used in some cases (13, 17, 18).

The US image of lipoma and angiomyolipoma of the liver shows the hyperechogenic structure with the homogenous interior and smooth contours (10, 11). This is caused by substantial damping of the ultrasound wave by fat and infiltrating character of the tumour growth. Therefore, the US angiomyolipoma image is often misdiagnosed as haemangioma (7). At this stage of diagnostic procedures patients are usually sent for CT, particularly that in the majority of them US examination discloses nodular lesions in the kidneys, which is always an indication for CT (5, 7, 13).

The CT image of fatty tumours in the liver is very characteristic – both lipoma and angiomyolipoma show the presence of negative density areas, below (-)20 HU (1, 10); the differences are found in the postcontrast phase: lipoma does not show visible enhancement while angiomyolipoma contains the point or linear foci enhanced in the arterial phase and visible in the parenchymal phase. The typical picture of angiomyolipoma was demonstrated in all our cases where the focal changes exceeded 5 mm. In smaller foci, even with the collimation 2.5 or 1.2 mm, it is difficult to obtain reliable measurements of density determining postcontrast enhancement, thus to differentiate small foci of lipoma from those of angiomyolipoma.

An extremely rare case of cystic teratoma containing the fatty tissue can be easily differentiated due to typical calcifications (13).

In agreement with other reports, multiple angiomyolipomas in the liver coexisted with analogical renal tumours containing fatty tissue.

In general, angiomyolipoma of the kidney is a single, small tumour of typical US and CT pictures, which usually do not require verification (5). The final diagnosis in atypical cases with low content of fatty tissue in the tumour is based on the biopsy with the confirmed presence of HMB-45 positive myoid cells (17). In the multiple form numerous, slowly growing tumours containing fatty tissue and vascular elements are observed. In the majority of cases, angiomyolipomas of the liver and kidneys have not tendency to malignant transformation. Nevertheless, one case of malignant AML of the liver and at least 7 cases of malignant form of AML of the kidneys were described (2, 15).

The majority of patients with AML of the kidney are treated conservatively, surgical treatment is undertaken only in tumours characterized by atypical CT image or those causing significant compression of the adjacent structures, which was the case in two of our patients (1, 5, 8).

In all three patients MSCT enabled to diagnose additional coexisting lesions. In the supradiaphragmatic region of the lungs small cysts surrounded by normal parenchyma were demonstrated characteristic of lymphangiomyomatosis (LAM) or tuberous sclerosis complex (TSC). Moreover, CT of the patient with periventricular nodules of the brain detected earlier showed pneumothorax. The recent reports show that 25–35% of women with TSC have pulmonary cysts; they may also have chylothorax and intraparenchymal nodules.

The radiological picture and histopathological changes in LAM may be very similar to those in TSC; and coexisting angiomyolipoma of the kidneys is observed in about 50% of cases (6).

In two patients described, including one with TSC, the CT scans also showed multiple osteosclerotic foci which occurred mainly in the spine. Such lesions have already been described in one TSC case by Pui et al. (14). There are no reports about sclerotic lesions in the spine coexisting with LAM, however, the symptoms of decreased bone density have been reported (16). Furthermore, CT fatty tumours in the region of the heart apex and ventricular septum found in one patient are worth stressing.

The cases presented by us indicate that CT-detected focal lesions containing fatty tissue located in the liver or kidneys are likely to coexist with other lesions, seemingly unrelated with the underlying disease but affecting the final diagnosis and management.

REFRENCES

1. Ahmadi T. et al.: Angiomyolipoma of the liver: significance of CT and MR dynamic study. Abdom. Imaging, 23, 520, 1998.
2. Dalle I. et al.: Malignant angiomyolipoma of the liver: a hitherto unreported variant. Histopathology, 36, 443, 2000.
3. Finegold M. J.: Tumors of the liver. Semin. Liv. Dis., 14, 270, 1994.
4. Fricke B. L. et al.: Frequency and imaging appearance of hepatic angiomyolipomas in pediatric and adult patients with tuberous sclerosis. AJR Am. J. Roentgenol., 182, 1027, 2004.
5. Gołębiowski M. et al.: Obustronny naczyniakomięśniakotłuszczak nerek u chorej ze stwardnieniem guzowatym. Pol. Przegl. Radiol., 62, 235, 1997.
6. Hansell D. M. et al.: Imaging of the Diseases of the Chest. Mosby, 680, 2005.
7. Hartleb M. et al.: Mnogie tłuszczaki wątroby. Hepatol. Pol., 3, 233, 1996.
8. Jover J. M. et al.: Lipoma of the liver associated with hepatocellular carcinoma and polycystic liver disease. Dig. Surg., 18, 323, 2001.
9. Kreel L. et al.: The radiological diagnosis and management of angiomylipoma of tuberous sclerosis. Australas. Radiol., 33, 176, 1989.
10. Langsteger W. et al.: Lipoma of the liver: computed tomographic, ultrasonographic and cytologic findings. Scand. J. Gastroenterol., 25, 302, 1990.
11. Linton P. L. et al.: Angiomyolipoma of the liver: immunohistochemical study of a case. Liver, 11, 158, 1991.
12. Nonomura A. et al.: Multiple angiomyolipoma of the liver. J. Clin. Gastroenterol., 20, 248, 1995.
13. Prasad S. R. et al.: Fat-containing lesions of the liver: radiologic-pathologic correlation. Radiographics, 25, 321, 2005.
14. Pui M. H. et al.: Bone changes in tuberous sclerosis mimicking metastases. Australas. Radiol., 40, 77, 1996.
15. Takahashi N. et al.: Malignant transformation of renal angiomyolipoma. Int. J. Urol., 10, 271, 2003.
16. Taveira-DaSilva A. M. et al.: Bone mineral density in lymphangioleiomyomatosis. Am. J. Respir. Crit. Care Med., 171, 61, 2005.
17. Tsui W. M. et al.: Hepatic angiomyolipoma: a clinicopathologic study of 30 cases and delineation of unusual morphologic variants. Am. J. Surg. Pathol., 23, 34, 1999.
18. Valls C. et al.: Fat in the liver: diagnosis and characterization. Eur. Radiol., 16, 2292, 2006.

SUMMARY

Multiple angiomyolipoma in the liver and kidneys are rare and may cause diagnostic difficulties. The authors present three cases diagnosed by US and MSCT. In all the patients the characteristic changes in the liver and kidneys were accompanied by cystic lesions in the lungs, in two cases – osteosclerotic changes were found in the spine; lipoma of the heart, retroperitoneal space, integuments

and changes in the central nervous system typical of tuberous sclerosis complex were also observed (each in one case). MSCT enables accurate and comprehensive diagnostic procedure in cases of fatty tumours of the liver and kidneys.

Mnogie angiomiolipoma wątroby i nerek diagnozowane metodą wielorzędowej tomografii komputerowej – opis przypadków

Mnogie angiomiolipoma wątroby i nerek występują rzadko i niekiedy mogą sprawiać trudności diagnostyczne. Autorzy przedstawiają opis trzech przypadków diagnozowanych w USG i metodą wielorzędowej tomografii komputerowej. U wszystkich pacjentek, poza charakterystycznymi zmianami w nerkach i wątrobie, stwierdzono współistniejące *cystic lesions* w płucach, w dwóch przypadkach występowały zmiany osteosklerotyczne kręgosłupa, stwierdzono po jednym przypadku tłuszczaka serca, powłok i zmiany w centralnym układzie nerwowym, typowe dla *tuberous sclerosis complex*. Tomografia wielorzędowa wykonana metodą wielofazową pozwala na dokładną i kompleksową diagnostykę w przypadku tłuszczowych guzów wątroby i nerek.