SUMMARY

Introduction
Carney triad is a rare non-hereditary condition characterized by gastrointestinal stromal tumors – intramural mesenchymal tumors of the gastrointestinal tract with neuronal or neural crest cell origin, pulmonary chondromas, and extra-adrenal paragangliomas. The term incomplete Carney triad more precisely refers to the occurrence of at least two of these tumor types. Carney triad named after J. Aidan Carney is considered to be a specific type of multiple endocrine neoplasia. Less than 30 cases of complete and less than 100 cases of incomplete Carney triad have been reported worldwide. Carney triad primarily affects young women (mean age of onset of 20 years).

Case outline
A 35-year-old female patient had an initial presentation at the National PET Center, Clinical Center of Serbia, after the gastrectomy, with multiple hepatic metastases as well as bilateral pulmonary chondromas. 18F-FDG PET/CT scan revealed the following: 1) intense 18F-FDG uptake in the liver metastatic lesions, with reduced physiological activity in the brain and heart, bowel, and renal tracer uptakes commonly known as FDG hepatic superscan; 2) multiple irregular-shaped lesions, mostly calcified in bilateral pulmonary parenchyma; 3) a nodular lesion in the left adrenal gland with accumulation of 18F-FDG in its anterior part.

Conclusion
The present study describes a hepatic superscan in a patient with incomplete Carney triad, including gastrointestinal stromal tumors and pulmonary bilateral chondromas, as well as a tumor in the left adrenal gland.

Keywords: GIST; 18F-FDG PET/CT; hepatic superscan; Carney triad; tartrate-resistant acid phosphatase

CASE REPORT

A 35-year-old female patient had an initial presentation at the National PET Center, Clinical Center of Serbia. Her medical history revealed a GIST diagnosed in 1989 and followed up with surgery (subtotal gastrectomy at the age of eight years, as well as total gastrectomy at the age of 23 years). The disease worsened in spite of surgery and chemotherapy and a recent clinical examination showed multiple hepatic metastases and bilateral pulmonary chondromas.

After the patient’s fasting six hours before the PET/CT study, and the median cubital vein cannulation, injection dose of 200 MBq 18F-FDG was applied, followed by a 90-minute data acquisition. 18F-FDG PET/CT examination on a 64-slice hybrid PET/CT scanner (Biograph; Siemens Medical Solutions USA, Inc., Malvern, PA, USA) was performed 90 minutes after tracer application. A three-dimensional PET scan (three minutes per bed position) and low-dose non-enhanced CT scan was acquired from the base of the skull to the mid-thigh. Multidetector CT was acquired with 120 kV and with automatic, real-time dose modulation amperage [CareDose4D (Siemens Healthcare GmbH, Germany)].

We report the visualization of an incomplete Carney triad in 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography – computed tomography (PET/CT) study in GIST postoperative phase and try to understand the anticipated coexpression of tartrate-resistant acid phosphatase in different organs, as they are liver and lungs.
Erlangen, Germany], with the baseline being 45 mA (slice thickness of 5 mm, the pitch of 1.5, and a rotation time of 0.5 s). CT, PET (attenuation-corrected), and combined PET/CT images were displayed for analysis on a single Multimodality Workplace (Siemens Healthcare GmbH).

The study revealed elevated right semi-diaphragm with heart dislocation to the left hemithorax (Figure 1).

There are multiple irregular-shaped lesions in bilateral pulmonary parenchyma, mostly calcified, partially consisting of a soft component, without increased uptake of 18F-FDG: 1) a single calcified (616 HU) lesion with a 13 × 12 × 16 mm diameter (AP × LL × KK) in the apical segment of the upper lobe of the right lung; 2) calcified (814 HU) lesion, soft tissue (57 HU), lesion diameter 36 × 33 × 33 mm (AP × LL × KK), perivascular paratracheal right at the Th3/Th4 level; 3) a single calcified lesion (540 HU), soft tissue (44 HU), lesion diameter 25 × 25 × 41 mm (AP × LL × KK) in the anterior segment of the upper lobe of the right lung; 4) a single calcified lesion (831 HU), soft tissue (71 HU), lesion diameter 35 × 30 × 23 mm (AP × LL × KK) in a lateral segment of the middle lobe of the right lung; 5) a single calcified lesion (652 HU), soft tissue (57 HU), lesion diameter 20 × 18 × 18 mm (AP × LL × KK) in the anterobasal segment of the low lobe of the left lung (Figure 2).

Augmented liver 20 × 24 × 27 cm (AP × LL × KK) contained multiple single and confluent hypodense lesions with intense uptake of 18F-FDG (SUVmax 27) and lesions without any uptake of 18F-FDG reflecting areas of necrosis in the “hepatic superscan” (Figure 1c, Figure 3).

There was a nodular lesion 30 × 12 mm in size (AP × LL) with intense accumulation of 18F-FDG (SUVmax 21) near the medial contour of the anterior part of the spleen (Figure 4).

The right kidney was dislocated caudally (level L3/L5) (Figure 3B). There was a nodular lesion diameter 31 × 16 × 21 mm (AP × LL × KK) in the left adrenal gland with accumulation 18F-FDG (SUVmax 3.1), in the anterior part of the nodular lesion (Figure 5).

Diffuse intense 18F-FDG uptake in the liver on PET, with reduced physiological activity in the brain and heart, bowel, and renal tracer uptakes is commonly known as FDG hepatic superscan (Figure 1c) [10, 11].

---

**Figure 1.** A – axial CT, fused PET/CT; B – coronal CT, fused PET/CT; C – (maximum intensity projection) PET images of elevated right semi-diaphragm with heart dislocation to the left hemithorax

**Figure 2.** A – coronal, B – sagittal, C – axial CT, fused PET/CT (mediastinal window) images of bilateral pulmonary chondromas

**Figure 3.** A – Coronal, B – Sagittal, C – axial CT, fused PET/CT images of multiple single and confluent hypodense lesions with intense uptake of 18F-FDG; lesions without any uptake of 18F-FDG show areas of necrosis in the hepatic superscan image
DISCUSSION

Multiple neoplasia syndromes are often considered with the presentation of multiple rare primary tumors in young patients. It is important to recognize the possibility of other primary tumors when associated neoplasms are detected [12]. The term Carney triad refers to the occurrence of at least two of the following tumor types: GIST, pulmonary chondroma, extra-adrenal paraganglioma. In a small percentage of affected patients, adrenocortical adenoma (a benign tumor of the adrenal gland) or esophageal leiomyoma (a benign tumor of the esophagus) may also occur [13]. The Carney triad is an extremely rare syndrome, with fewer than 30 cases reported with all three tumors present, and fewer than 100 incomplete cases having two of the three tumor types present [13]. According to Carney in 1999, chondromas developed in 76% of patients [13].

The present report describes the patient with incomplete Carney triad, including GIST and pulmonary bilateral pulmonary chondromas, as well as the tumor in the left adrenal gland. This is a demonstration of the 18F-FDG PET/CT utility in diagnosis / differential diagnosis in some of the rare diseases. The hepatic superscan was demonstrated in the reported case of an incomplete Carney triad.

The multiorgan (liver, lungs) molecular coexpression of tartrate-resistant acid phosphatase (TRAP) in immunocytes belonging to monocyte/macrophage lineage should be anticipated as of importance in the pathogenesis of this clinical case. Physiologically, TRAP is primarily a cytochemical marker of macrophages, osteoclasts, and dendritic cells [14]. Under normal circumstances, TRAP is highly expressed by osteoclasts, activated macrophages, neurons, and by the porcine endometrium during pregnancy [15, 16]. In newborn rats, TRAP is also detectable in the spleen, thymus, liver, kidneys, skin, lung, and heart at low levels. TRAP expression is increased in certain pathological conditions. These include leukemic reticuloendotheliosis (hairy cell leukemia), Gaucher’s disease, HIV-induced encephalopathy, osteoclastoma and osteoporosis, and metabolic bone diseases (available at https://en.wikipedia.org/wiki/Tartrate-resistant_acid_phosphatase).

TRAP is a glycosylated monomeric metalloprotein enzyme expressed in mammals and characteristic for its expression in activated osteoclasts and macrophages and was proposed as a driver of metastasis and was associated with clinically relevant parameters of cancer progression and cancer aggressiveness [17, 18].

The coexistence of an adrenal tumor with incomplete Carney triad indicates possible neuroendocrine origin and inclusion in multiple endocrine neoplasia syndromes.

Conflict of interest: None declared.
REFERENCES

3. Genetic and Rare Diseases Information Center (GARD) [Internet]. Gaithersburg, MD, USA. Carney Triad [cited 2019 Sep 05]; [about 5 screens]. Available from: https://rarediseases.info.nih.gov/diseases/10924/carney-triad.

Суперскен јетре 18F-FDG PET/CT у некомплетној Карнијевој тријади

Љиљана Зивгаревић, Небојша Козаревић, Светлана Жунић

Клинички центар Србије, Центар за нуклеарну медицину, Национални ПЕТ центар, Београд, Србија

САЖЕТАК

Увод Карнијева тријада је ретко ненаследно обољење које се карактерише присуством гастроинтестиналног стромалног тумора – интрамуралног мезенхималног тумора гастроинтестиналног тракта пореклом неуралног гребена, присуством хондрома у плућним ткивима и экстраадреналних параганглиома. Периодични обилази у кругу пацијенте, присуство тумора у левој надбубрежној жлезди. Фокусира се на присуство следећег: 1. Калицификоване, са мањим уделом мекоткивне компоненте, вишеструке лезије неправилног облика, највећим делом налазе се у јетри и обостраним плућним хондромима. 2. Суперскен јетре, са редукованим везивањем у мозгу, срцу, цревима и присуство тумора у левој надбубрежној жлезди. 3. Картине тумора у метастазама у јетри и обостраним плућним хондромима. Студијом 18F-FDG PET/CT доказано је присуство следећег: 1. Интензивно везивање 18F-FDG у бројним метастазама у јетри, са редукованим везивањем у мозгу, срцу, цревима и бубрезима, што одговара опису суперскена јетре; 2. Виештруке лезије неправилног облика, највећим делом каплетане, са мањим уделом нокотивих компоненте. 3. Картине тумора у метастазама у јетри и обостраним плућним хондромима. 4. Картине тумора у метастазама у плућима, као и присуство тумора у левој надбубрежној жлезди. 5. Картине тумора у предњем делу појачано накупија 18-FDG.

Закључак Овај приказ опишује налаз суперскена јетре код болеснице са некомплетном Карнијевом тријадом, која у конкретном случају укључује желудачни гастроинтестинални стромални тумор и обострани хондром у плућима, као и присуство тумора у левој надбубрежној жлезди.

Кључне речи: ГИСТ; 18F-FDG PET/CT; суперскен јетре; Карнијева тријада; кисела фосфатаза резистентна на тартрат