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Letter to the Editor

Validation of giant cell arteritis diagnosis code in the French hospital electronic database

Giant cell arteritis (GCA) is a rare form of large-vessel vasculitis which predominantly affects women over 50 years of age [1]. GCA usually involves granulomatous inflammation of the aorta and its major branches, with a predilection for extracranial branches of the carotid artery. The annual incidence rate per 100,000 inhabitants aged ≥50 years is between 5.8 and 20 in European countries [2].

Population-based studies using health care or insurance databases are very useful to assess the epidemiology of rare diseases such as GCA [2,3]. However, health insurance databases are primarily compiled for reimbursement purposes and not research. Consequently, there is still uncertainty about the accuracy of disease and event coding in these databases.

The French Health Insurance System Database called SNDS (*Système national des données de santé*) virtually covers the entire French population. It links administrative, out-patient health care dispensing and in-patient data. The latter consists in the PMSI (*Programme de Médicalisation des Systèmes d'Informations*) hospital database. It records data relating to all hospital stays in both private and public hospitals, including admission and discharge dates, one primary diagnosis code ± one related diagnosis code (generally corresponding to the reason for hospitalization), and unlimited associated diagnosis codes (corresponding to comorbidities or to events that occurred during the stay), procedures, and costly drug-dispensing data. Diagnoses are encoded at discharge using the International Classification of Diseases, 10th version (ICD-10th) by the physician in charge of the patients or by professional coding technicians reviewing the medical records.

The aim of this study was to assess the accuracy of GCA diagnosis codes [M31.5 – "GCA with polymyalgia rheumatica (PMR)" and M31.6 – "other GCA"] in the PMSI. We assessed the positive predictive value (PPV) (whether M31.5 or M31.6 codes indicate a hospital stay for GCA) and sensitivity (whether hospital stays for GCA are accurately encoded with the M31.5 or M31.6 codes).

We extracted all hospital stays with M31.5 and/or M31.6 codes as primary, related or associated discharge diagnosis from the 2013, 2014 and 2015 PMSI data recorded at Toulouse University Hospital (South of France, 2880 beds), reviewed by two GCA experts (LC and GP) to assess whether the patients had GCA or GCA with PMR.

Patients were first considered as having a GCA when at least 3 of the following criteria were met, based on the 1990 American College of Rheumatology (ACR) classification criteria [4]: (1) age at onset ≥50 years, (2) new headache (new onset or new type of localized head pain), (3) temporal artery abnormality (tenderness on palpation or decreased pulsation, unrelated to arteriosclerosis of the cervical arteries), (4) elevated erythrocyte sedimentation rate (ESR ≥ 50 mm/h) and (5) temporal artery biopsy (TAB) consistent with GCA (biopsy specimen with artery portraying biopsy vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells). Further, if criteria (5) was not met, an arterial imaging consistent with GCA was mandatory to retain GCA diagnosis: (1) highly suggestive angio-MRI (magnetic resonance imaging) or –CT scan (computed tomography): thickened and contrast-enhanced arteries in T1- weighted sequences, especially aorta (> 3 mm) and epi-aortic arteries (> 1 mm); (2) highly suggestive positron emission tomography (PET) with high ^{18}FDG uptake scan of large artery walls, especially the aorta and epi-aortic arteries; (3) highly suggestive Doppler ultrasound: 1 mm thick, circumferential and diffuse hypo-echoic halo ± occlusion ± stenosis of temporal or axillary arteries.

The diagnosis of associated PMR was confirmed when at least a 4 points overall were recorded in relation to the following criteria, based on the 2012 EULAR/ACR classification criteria [5] for this disease: (1) morning stiffness > 45 min (2 points), (2) hip pain or limited range of motion (1 point), (3) normal rheumatoid factor or anti-citrullinated protein antibodies (2 points) and (4) absence of other joint pain (1 point).

The PPVs and their 95% confidence intervals (CI) were calculated for all hospital stays and then stratified according to the category of diagnosis code (primary, related, and associated). The patients' source for assessing the sensitivity was the Toulouse GEFA registry (*Groupe d'Etude Français des Artérites des gros vaisseaux*). This registry aimed at following up all GCA patients diagnosed in the Internal Medicine Department at Toulouse University Hospital since 2012. We identified all patients included in the GEFA, diagnosed with GCA between 2013 and 2015 and who were admitted at Toulouse University Hospital with disease onset. Then we assessed whether their hospital stays were encoded using the M31.5 or M31.6 code in the PMSI.

Between 2013 and 2015, 507 hospital stays were identified with M31.5 and/or M31.6 codes as primary, related or associated diagnosis, corresponding to 302 patients (median age: 79 years, females: 68.5%). All of them had the wording of GCA disease in their medical charts.

Among these 302 patients, we could check the GCA classification criteria for 170 patients, 160 of whom met those criteria. 100% (160/160) were over the age of 50, 82.1% (129/157) had a new headache, 62.1% (90/145) a positive TAB, 31% (45/145) an abnormal induration temporal artery, 94.4% (151/160) an ESR ≥ 50 mm/h and 81.8% (99/121) had a positive MRI, CT, PET scan or Doppler. Overall, 160 patients were true positives. The PPV was 94.1% (95% CI: 89.5–96.7).

When stratifying by diagnosis categories, higher PPVs were recorded for primary and related diagnoses than for associated diagnoses (Table 1), with 10 false positive cases (PMR without GCA for 8 patients, rheumatoid polyarthritis and ANCA-associated vasculitis, 1 patient each).

During the same period, 84 newly diagnosed GCA patients have been recorded in the GEFA registry database. The initial hospital stay was encoded M31.5 and/or M31.6 for 81 out of these patients, all of them coded as primary diagnosis. Consequently, the sensitivity of the GCA diagnosis code in the PMSI database was estimated at 96.4% (95% CI: 90.0 to 98.8%), 3 false negative cases coded M35.3 (PMR without

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Table 1
Positive predictive values of ICD-10th GCA code (M31.5 or M31.6) in the SMDS/PMSI database.

Category of discharge diagnosis code	Number of patients with M31.5 or M31.6 codes	Number of true positive cases	PPVs (95% CI)
Overall	170	160	94.1% (89.5–96.7)
Principal	99	96	97.0% (93.6–100)
Related	22	22	100%
Associated	47	42	89.4%(80.6–98.2)

GCA).

Of the 17 patients with a hospital stay coded M31.5 as primary, related or associated diagnosis, all of them had the wording GCA and PMR disease in their medical charts. Thirteen met GCA and PMR diagnosis criteria after medical chart review. The overall PPV was 76.5%. (95% CI: 52.7 to 90.4%). Four false positive cases were attributed for 3 patients to PMR without GCA and for 1 patient to unclassified arthritis. Among the 18 newly diagnosed cases of GCA associated with PMR recorded in the GEFA registry during the study period, the initial hospital stay was encoded M31.5 in 16 cases. Consequently, the sensitivity was 88.9% (95% CI: 67.2 to 96.9), 2 false negative cases coded M35.3 (PMR without GCA).

To the best of our knowledge, this is the first study to assess the accuracy of ICD-10th GCA diagnosis codes in an electronic hospital database. This study demonstrated excellent PPVs for the M31.5 and M31.6 codes of primary and related diagnoses in the Toulouse University Hospital database. The sensitivity was also very good. Only one other study has investigated the accuracy of GCA diagnosis codes in another hospital database, the Hospital Episode Statistics database linked with ambulatory care data in the Clinical Practice Research database (CPRD). In this database, the Oxford medical information system is used for diagnosis coding, which is based on the ICD-9th [6]. Hospital discharge summaries were requested in a sample of 50 cases with a diagnosis of GCA identified in the CPRD. Of the 50 requested, 45 (90%) were retrieved. The extent of investigation of patients varied and was insufficient to allow strict diagnostic criteria application. Overall, on the basis of typical symptoms and a clinical response to corticosteroids, the physician diagnosis of GCA was supported in 41/45 (91%) cases [7].

Some limitations of our study must be acknowledged. First, this study was conducted in a single university center and may not reflect the performance of diagnosis coding at the national level. However, coding is similar in all private and public hospitals due to application of validated national standards [8]. Furthermore, demographic data of patients included in this study is in accordance with French national and international cohorts [9,10].

Overall, this validation study suggests good reliability of the ICD-10th GCA diagnosis code in the French hospital database.

Conflict of interest statement

All authors declare: no support from any organization for the submitted work; no financial relationships with any organizations that might have an interest in the submitted work in the previous 3 years; no other relationships or activities that could appear to have influenced the submitted work.

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